PROGRESSIVE

MUSCULAR DYSTROPHY.

Comparisons of some of the varieties with each other,

AND WITH

Progressive Muscular Atrophy.

Twenty-six Original Photographs.

BY

J. H. CROCKER, M.B.,

Bachelor of Surgery, Victoria University; Licentiate of the Royal College of Physicians, London; Member of the Royal College of Surgeons, England: etc.

MANCHESTER:

ABEL HEYWOOD & SON, 56 AND 58, OLDHAM STREET.

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MUSCULAR ATROPHY OF MYOPATHIC ORIGIN.

DURING the past half-century many varieties of Muscular Atrophies have been recorded, now supposed to be of Myopathic origin—*i.e.*, not due to any lesion of the spinal cord or nerves, but to a defect of the muscles themselves—but until a few years ago the chief attention was given to that form characterised by overgrowth of the connective or fatty tissue in some muscles in connection with atrophy of the muscular fibres, known as Pseudo-hypertrophic Muscular Paralysis. In 1884 Erb described a form of atrophy coming on about puberty, designated Erb's Juvenile Form of Idiopathic Muscular Atrophy, and since then more careful study has been made of the various types of this affection.

Among the cases and varieties of Myopathic Atrophies that have been published may be mentioned the following.

(*a*) Dr. Meryon,[1] in 1852, described an hereditary affection in several boys of two different families under the heading of "Granular and Fatty Degeneration of the Voluntary Muscles." Charcot and Sir Wm. Roberts thought they were due to progressive muscular atrophy, Duchenne and Friedreich looked upon them as allied to pseudo-hypertrophic paralysis, while Gowers[2] is of opinion they were the latter complaint with the enlargement not conspicuous.

(*b*) Duchenne, in 1855, described an "infantile form" of muscular atrophy which began in the face at an early age,—about five to seven years,—and affected the upper extremities later,—about

(1) "On Granular and Fatty Degeneration of the Voluntary Muscles," Medical Chirug. Trans., 1852—*B.M.J.*, 1870.

(2) "Diseases of the Nervous System," 1892, Vol. 1, p 506; also appendix to his lecture on P. H. P., 1879.

the eleventh or twelfth year. Generally, advice was not sought till the latter symptoms developed. Later still, other muscles were affected. A curious coincidence is that the father of the first two cases recorded died at forty-eight, of Cruveilhier's palsy, and that his father also died of the same complaint.[3] Although rare, down to 1872 Duchenne had observed twenty cases of this ailment. In 1874 Landouzy and Déjérine published several cases, and one in 1885 with post-mortem reports. The description of these cases shows that they were of the Facio-Scapulo-Humeral type, in England often spoken of as the Landouzy-Déjérine type, but Erb in his article speaks of it as "Duchenne's Infantile form."

(*c*) Erb,[4] in 1884, described a form under the title of "Dystrophia Musculorum Progressiva Juvenilis." He looks on this as the simplest form of muscular atrophy, it having the fewest complications and the simplest course. His definition of it is: chronic advancing atrophy and weakness of numerous voluntary muscles—beginning generally in youth, rarer in middle age and frequently upon a family basis—with characteristic localisation of the disease in the upper half of the body, which is generally earlier and more severely affected, especially the *pectorals*, *latissimus dorsi*, *trapezius*, *rhomboids*, and other scapular muscles, the flexors of the upper arm, including the *supinator longus*, then attacking the *triceps*. The forearm and hand muscles remain unaffected but not invariably in advanced cases. In the lower half of the body, chiefly the extensors of the trunk, part of the abdominal muscles, glutei, thigh muscles and finally calf and peroneal region. Hypertrophy may appear in some of the muscles—partly true and partly false. In the end, scarcely one muscle may remain unaffected in the body. In consequence of the atrophy, peculiar displacement of the scapulæ, lordosis, peculiarities in gait, etc., result.

(*d*) In 1886, Dr. Tooth[5] described some cases of Peroneal Atrophy, the majority of these being probably due to Neuritis, or

(3) Fagge's "Medicine," 1891, Vol. 1., p 513.

(4) Deutsches Archiv.f.klin. Med. : 1884.

(5) The Peroneal Type of Progressive Muscular Atrophy—Lewis, 1886.

of spinal origin (R.D. Fibrillar twitchings, etc.) but some may have been of myopathic origin.

(*e*) Leyden and Moebius have described an hereditary muscular atrophy beginning in the lower extremity, calves generally, ascending to the thigh and lumbar region, and only after years to the upper extremity.

(*f*) Primary Multiple Muscular Atrophy, described by Dr. Poore in his edition of Duchenne's work; supposed by Gowers to be Pseudo-hypertrophic Paralysis without the enlargements.

(*g*) Duchenne, in 1861,[6] described an affection characterised by enlargement of certain muscles, giving the patient the appearance in some parts of an exaggerated "Farnese Hercules," offering a strong contrast to the atrophy which is commonly found in the upper parts. He gave it the name of Pseudo-hypertrophic Muscular Paralysis.

Of recent years on the Continent the usually adopted classification for Myopathic Atrophies has been as follows,—

1. Duchenne's Infantile form of Idiopathic Muscular Atrophy.
2. Erb's Juvenile ,, ,,
3. Leyden's Hereditary ,, ,,
4. Pseudo-hypertrophic Muscular Paralysis.

In 1891 Erb,[7] published the results of his investigation in "Progressive Muscular Dystrophies," and as I am not aware that they have been given in English, I append a short resumé. He endeavours to show that among the various types which have been described as belonging to the primitive myopathies, there are so many points of resemblance, and transitional forms, that they are only different manifestations of one and the same disease. He gives a series of forty cases of the Juvenile and Pseudo-hypertrophic types, and proceeds to compare them, concluding as follows:—A careful consideration of all these observations, shows, as it seems to me in a convincing manner, how close and intimate are the relations between these two types, so close that in many cases any

(6) Electrisation Localisee, 1861.

(7) Deutsche Leitschrift fur Nervenheilkunde, April 30 and July 24, 1891.

differences between them disappear. In Pseudo-hypertrophic paralysis there is always a widely distributed atrophy of many muscles, especially in the upper half of the body, shoulder girdle, and upper arm; the localisation of the atrophy is always the same as in the Juvenile type; the differences which may exist between the Juvenile type and the Pseudo-hypertrophic are not greater than exist between the individual cases of Juvenile Atrophy themselves or of the individual cases of Pseudo-hypertrophy. The relations of the muscles are also alike as regards their response to mechanical and electrical irritation, and there are no fibrillar contractions in either as a rule. The disturbances in gait, movements, etc., are identical.

He next gives a short account of ten cases of Duchenne's Infantile form (Landouzy—Déjérine) where there is primary implication of the face, ending with remarks upon its relations to the first two types. His observations upon these points are almost a repetition of what he has already said of the first two.

Then follow his remarks upon Leyden's hereditary form and what he calls Indefinite types, that is, cases where even a skilled observer would find it difficult to ascribe it to any of the four. He gives one such example. He also remarks that in the same family there may be found several of the types.

Marie and Guinon,[8] have reported on the similarity of Idiopathic Muscular Atrophy and Pseudo-hypertrophic Muscular Paralysis and gave details of some cases at the Salpêtrière.

From perusal of the clinical reports of the various cases, one is struck with the sameness of the affected regions: a case, which may be in one class or type at one time, may in a few years be fairly considered as belonging to another. The great majority are what we should call Scapulo-Humeral, sometimes varied with implication of the face—and Erb says the face muscles are affected more frequently than is generally supposed—at other times with more or less atrophy or hypertrophy. According to Duchenne, in the earlier cases the face is more likely to be affected, the reason for which is difficult to explain, as is also the fact, that the

(8) Revue de Med. 1885.

Pseudo-hypertrophic form affects boys more than girls, and as a rule commences earlier than the other forms.

The following nine cases of simple Idiopathic Muscular Atrophy, and two of Pseudo-hypertrophic Muscular Paralysis are interesting, inasmuch as they present different types, and variations in the same type, well illustrating many of the features referred to by Erb.

SIMPLE IDIOPATHIC MUSCULAR ATROPHY.

MOTHER AND SEVEN CHILDREN DIFFERENTLY AFFECTED.

Case 1.—Face only affected. — Mrs. P——, aged 54, mother of the following seven cases, has always enjoyed good health; did not know anything was the matter with her. She has three sons and five daughters alive; none dead. No history of a similar affection in antecedents or present relatives, with the exception of the above.

Present Condition, January, 10th, 1894.—She looks in very good health and apparently nothing is wrong with her.

Face.—The muscles affected are the *Frontales*, *Corrugator Supercilii*, *Orbicularis Palpebrarii* and *Orbicularis Oris*. The *Zygomatics*, which are said to be the first affected generally, are slightly impaired. The other facial muscles are normal, and there are no muscles affected on the body or extremities.

Facial Movements.—In consequence of the affected muscles she cannot frown, and there is a faint movement only on attempting to wrinkle the forehead. In endeavouring to close the eyes, the lids do not completely approximate, about $\frac{1}{8}$ inch being left between the upper and lower margins. There is no wrinkling of skin or evidence of any movement of the *Orbicularis Palpebrarum*, either of Orbicular or Palpebral portion, the lid falls just as if due to relaxation of the *Levator Palpebræ Superioris*. She can laugh, but the angle of the mouth is only slightly elevated. She cannot say O, clearly, and cannot round the mouth as in whistling; the photograph No. 1 was taken when she was endeavouring to do this.

Remarks.—Until the disease was recognised in the children, it had not been noticed in this case. From the photograph one would not suspect any affection of the facial muscles, as the naso labial folds are well marked, and the Zygomatics act. It is interesting to note that the Zygomatics were not effected in a case recorded in 1893.[9] Evidently this case has not advanced much during the past eleven months, as the symptoms are very little worse than when Dr. Williamson examined her.[10]

It is evident that occasionally these cases may remain stationary or advance very slowly, thus resembling Pseudo-hypertrophic cases. Gowers, in the appendix to "Pseudo-hypertrophic Muscular Paralysis," mentions the case of Rev. S. S., aged 74, he always had conspicuously large calves, but suffered no inconvenience, and was always able to take active exercise. He had seven or eight children, of whom two sons and one daughter developed Pseudo-hypertrophic Muscular Paralysis.

Case 2.—Face, shoulders and arms affected.—Mary P——, age 23, daughter of case 1, single, mill hand.

Past History.—Was well up to ten years of age, when she began to get thinner, and her chest seemed to sink in. She had a fit at the age of 16, and about 12 months after she noticed a growing weakness in the shoulders and arms, she has gradually gone worse since, but still goes to work at the mill. She has had no fits for 12 months. Is not aware when the face was first affected.

Present Condition.—The patient looks thin, and has the characteristic "Mask-like" or "Myopathic Face," with "Tapir-like" mouth as described by Landouzy and Déjérine. Dull, expressionless, and muddy complexion.

Face.—All the muscles are affected, some completely. The muscles of the eyes intrinsic and extrinsic, as well as those of the tongue, palate, pharynx and larynx are unimpaired.

Neck.—*Trapezius*—completely atrophied at the lower and much wasted at the upper part, especially right side. *Levator anguli*

(9) *Deutsche Leitschrift*, October, 1893.

(10) *Medical Chronicle*, September, 1893.

scapulæ somewhat atrophied, especially on right side. *Sterno cleido mastoid* very thin and narrow, clavicular portion much atrophied.

Shoulders and Arms.—Appearance—The peculiar wing-like position of the scapulæ is very marked, the vertebral border being tilted towards the horizontal, whilst the inferior angle points backwards, downwards, and inwards, especially when the weight of the arms is thrown on the muscles. The *clavicle* is depressed at its outer end, and the *coracoid process* is very distinct.

Muscles.—The *Rhomboids* are completely atrophied. See photo No. 4. *Serratus Magnus* atrophied on both sides. *Pectoralis major and minor* have disappeared, excepting a few fibres at the upper part of major. *Teres major and minor* atrophied. *Supraspinatus* wasted, but not completely. *Infraspinatus*, the thickest muscle by far on each side, is hypertrophied about the middle, and to the touch feels hard and bullet-like. See photo No. 4. *Deltoids*—decidedly atrophied in upper part, but lower half on each side is enlarged and feels very hard, similar to that of pseudo-hypertrophic paralysis, most marked on the right side. *Latissimus dorsi* atrophied.

Arm.—*Coraco brachialis*, *biceps*, *triceps*, and *supinator longus* atrophied somewhat on each side.

Forearm.—*Extensors carpi radialis longior* a little wasted. The remaining muscles of forearm and hand are normal on both sides.

The pelvic muscles and those of the lower extremities are not much wasted, and perform normal movements, except that the feet can only be brought voluntarily up to an angle of 45 degrees with the legs in dorsiflexion, and there is slight inversion of the left foot.

Movements.—Face.—Cannot wrinkle the brow or frown, nor quite close the lids by $\frac{1}{8}$th of inch. See photo No. 2. Cannot point the lips as for whistling. Can only raise the lips the least bit on the left side. Though impossible to elevate the outer angle of the mouth, she can draw it outwards.

Shoulders.—She shrugs the shoulders a little, and can rotate the scapulæ slightly.

Arm.—Abduction is very weak, chiefly done by rotation of the scapula, but she cannot raise the arms to a horizontal position—this shows weakness of the *Deltoids* in spite of enlargements. See photo No. 3. With the arms hanging down she can just bring the right one behind the back, but cannot do so with the left. She can only touch the opposite clavicle with the fingers after considerable effort.

Forearm.—Flexion and extension of the elbow very weak, and very little resistance arrests the action. Owing to the atrophy of the *Supinator longus* she cannot turn the handle of a door, and when tying a tape or fastening a button the action is very striking, the arm being semiflexed at the elbow and the supination and pronation movements done by rotation of the arm and forearm together, the elbow travelling through a large arc with each movement. The muscles of the forearm and hand are normal in their action on both sides.

Standing.—No abnormal curvature.

Sitting.—Can get up from a chair easily.

Walking.—No oscillation.

Electrical Reactions of the Muscles.—Diminished to both Faradic and galvanic currents. No reaction of degeneration. No fibrillar contractions.

Measurements.—Shoulder, 10½ inches, right; 10½ inches, left. Arm (middle), 6 inches, right; 6¼ inches, left. Forearm, 7 inches, right; 7 inches, left. Thigh (middle), 15 inches, right; 15 inches, left. Calf, 11 inches, right; 11 inches, left.

Reflexes.—*Organic*, Normal and unimpaired. *Superficial*, Plantars present. *Deep*, Knee jerk weak, but obtainable. No ankle clonus.

Sensation.—Normal.

Remarks.—This case is one of the Facio-Scapulo-Humeral, Duchenne Infantile, or Landouzy-Déjérine type, and is interesting, inasmuch as the Gluteal and Thigh muscles are very little affected in spite of the disease having existed for 12 years.

The condition of the Infraspinatus and Deltoid muscles, and absence of the Latissimus dorsi on each side, is shown in photo

No. 4, which is almost identical with the illustration 149 in "Gower's Diseases of Nervous System," Vol. I., 1892.

In its course in this patient, the disease is not so rapid as with the brother, who is in a worse condition, notwithstanding he only began three years ago. In speaking of Pseudo-hypertrophic paralysis, Gowers[11] states that when girls are affected the course is slower and not so severe.

Case 3.—Face and nearly all the voluntary muscles affected. Edward P——, aged 17, single, collier; brother to case 2. Was a delicate baby and did not walk until 2½ years of age, since then he has enjoyed good health up to three years ago, when he began to feel tired in his arms and weak in his back, so as to interfere with his occupation. Soon after he noticed his legs were weak when he walked far, and he dropped his toes a little. Eventually he gave up work owing to the weakness continuing. Does not know when the face was first affected.

Present Condition, January 10th, 1894.—The Myopathic face, with "Tapir-like" mouth, thick everted lips and dull expressionless countenance ~~are~~ well marked—see photos Nos. 5 and 6. Naso labial groove very faintly marked. There seems to be atrophy of all the muscles of the face, with the exception of a few fibres of the elevator of the upper lip on the left side, and a few elevating fibres in the lower lip.

The intrinsic and extrinsic muscles of the eye ball are normal, but like his mother and sister he cannot quite approximate the lids. The muscles of deglutition, etc., are not affected.

Neck.—*Trapezuis* wasted at lower part, not so much at upper. *Sterno-cleido-mastoid* much wasted especially clavicular portion on the right side. *Levator Anguli Scapulæ* not so much wasted. *Rhomboids* wasted.

Shoulder and Arm.—Depression of outer end of Clavicle, Coracoid process well shown. "Winging" and rotation of scapulæ,

(11) Gowers' Diseases of Nervous System, Vol. I., 1892. p. 506

vertebral border nearly horizontal, acromion downwards, and glenoid cavity at right angles to shaft of humerus, looking almost directly downwards. The upper arm is very thin. See Photos Nos. 5, 6, etc.

Muscles.—*Supraspinatus* a little atrophied. *Infraspinatus* hard to the touch and a little tendency to pseudo-hypertrophy. *Deltoid* much atrophied. *Latissimus dorsi and Serratus Magnus* apparently much wasted. *Pectorals* wasted. *Coraco-brachialis, Biceps, Triceps, and Supinator longus* all much atrophied, and contrast with the non-attenuated forearm. See photos Nos. 5 and 6. The forearm muscles and intrinsic muscles of the hands are now beginning to be a little affected.

Body.—Chest is thin and ribs easily defined. Abdominal wall thin and muscles distinctly atrophied. See Photos. Nos. 5 and 6. Muscles of back are thin.

Pelvis.—Rotated forwards and downwards on femora when standing.

Muscles.—The *Glutei* are much wasted, and the bony pelvis easily defined. Upon pressing into the iliac fossa there is marked atrophy of the *Iliacus*.

Thighs.—Atrophy most marked in the *Rectus femoris* and *Vasti* on each side, particularly the left.

Legs.—Dropping of feet showing *Peronei and Anterior Tibial* muscles are affected. *Calf muscles* are very slightly affected.

Movements.—*Face*, cannot frown, whistle, etc.

Shoulders.—Can shrug, but chiefly due to rotation of the Scapulæ by means of the remaining portion of the Trapezius, etc. Can rotate the arm, but abduction is difficult; cannot raise arm to horizontal position.

Arm.—Flexion and extension at elbow very weak.

Forearm, Supination.—Similar to sister, peculiar movements in fastening a button.

Thighs.—Flexion weak, extension weak, worst on left side.

Legs.—Cannot bring feet up to right angle with leg.

Position when standing.—Lordosis well marked, although

muscles of the back are weak, yet the chief cause is due to the tilting of the pelvis owing to the weak Glutei.

Sitting.—The lordosis disappears when the Pelvis is placed on the Tuberosities of the Ischii, compare photos, Nos. 5 and 6.

Walking.—The peculiar oscillating waddle, similar to that of Pseudo-hypertrophic cases.

Getting up from Recumbent Position. –Owing to the weakness of the extensors of the knee and hip, he has to assist them by an alteration of leverage which is explained later on when describing a Pseudo-hypertrophic case. After getting from the ground onto his feet with knees flexed, he throws the chief weight on to the right leg, and pushes the left leg out and back, then with hand on right knee, he extends that by the alteration of the position of the weight, and next he assists the weak Glutei by climbing up his thighs. See the five photos, Nos. 7, 8, 9, 10 and 11.

Measurements.—Shoulder, 11½ inches, right; 11 inches, left. Arm (middle), 5½ inches, right; 5½ inches, left. Forearm, 7 inches, right; 7½ inches, left. Thigh (middle), 13½ inches, right; 11½ inches, left; Calf, 12 inches, right; 12 inches, left.

Electrical Reaction.—Diminished to Faradic and Voltaic currents. No R. D.

Reflexes.—*Organic*: Normal. *Superficial*: Plantar and Cremasteric present. *Deep*: Knee jerk in right leg very feeble. No ankle clonus.

Sensation: Normal.

Remarks.—This case has many points worthy of consideration. Having the face affected it is like a case of Duchennes "Infantile" form: beginning in adolesence, and in other respects, it is like a case of Erb's "Juvenile" form. The extreme atrophy of nearly all the voluntary muscles, together with the emotionless face are very striking, as shown in the photographs, Nos. 5 and 6. There is complete disappearance of the *Latissimus dorsi*, and evidently even the abdominal muscles have atrophied to a great extent. The mode of walking, method of getting up from the recumbent position, the lordosis due to tilting of the pelvis which is shown by

the disappearance of the curve when sitting, are important points, as they are identical with that which has heretofore been considered characteristic of pseudo-hypertrophic muscular paralysis alone. The rapidity with which this case has progressed is remarkable, considering he left his work less than three years ago. See photos No. 5 to 11.

The foregoing three cases were examined by Dr. Williamson twelve months ago, and I have to thank him for kindly furnishing me with their address: When I examined them on January 10th, 1894, the mother assured me all the others were quite well and at work; however, on February 3rd, I journeyed to their home and waited until they all returned from work. I found, with the exception of one son, all had some muscles affected, chiefly confined to the face. The father of the family is a blacksmith. I have examined him and find nothing wrong; the son John P——, aged 20, works with his father, and has no apparent affection. The face muscles act normally, and all the muscles of the shoulder and arm are well developed, excepting the Infraspinatus.

The following five cases may have existed for some time unsuspected, they are interesting as showing the conditions in the early stage which is not often seen, on account of advice only being sought when weakness is pronounced.

Case 4.—Alice Ann P——, aged, 28, eldest of the family, sister to cases 2 and 3. Looks healthy and strong.

Muscles Affected.—Face: The *Frontales*, the *Orbicularis palpebrarii*, and *Orbicularis oris*.

Neck.—*Trapezius* thin and wasted below; all other muscles normal and strong.

Actions: She cannot frown or wrinkle the forehead. Upon closing the eyes, the lids only just approximate, and even with the greatest efforts of resistance on her part the lid is easily raised by the finger. The skin around the eyes wrinkles very little during this action. The *Zygomatics* act, but she cannot shorten the mouth, or use the Orbicular muscle, as for whistling. All movements of the body and extremities are normal and strong.

Case 5.—Ellen P——, age 25, sister to above cases. She is well and strong. Only the *Frontales*, *Corrugators supercilii* and *Orbicularis palpebrarii* very slightly affected. All other muscles of the face, body, and extremities are normal. She can slightly wrinkle her brow, but cannot frown; and the eyes can be closed, but the wrinkling around is not very marked. There is no impairment in laughing or whistling.

Case 6.—Elizabeth P——, age 16, sister to above cases. Face assuming the "Myopathic" characteristics.

Face.—Muscles Affected: *Frontales*, *Orbicularis palpebrarii*, *Orbicularis oris*, and *Zygomatics*.

Actions: Cannot frown or wrinkle the forehead. Lids just approximate but weak, and there is no wrinkling of the skin around the eyes. She has no power of shortening or shaping the mouth as for whistling. She can elongate the mouth, but cannot elevate the angles.

Neck.—Muscles Affected: *Trapezius* atrophied below and weak above. *Sterno cleido mastoid* thin, especially the clavicular portion. *Levator anguli scapulæ* present.

Shoulder and Arm.—Position of Scapulæ: Movements are very free, the scapulæ being slightly lower and further forwards than normal. The effect of the *Trapezius* being atrophied at the middle and lower part accounts for this, and also allows the *Rhomboideus minor* to show up on each side, where the inferior border forms an acute angle with the vertebræ owing to the insertion being so much lower than its origin. There is slight winging of the scapulæ. Another noticeable peculiarity resulting from the rotation forwards and winging of the scapulæ is that when the arms are held out horizontally, and in the transverse plane of the body, the head of the humerus is very prominent in front, and the *Deltoid* looks as if chiefly posterior instead of superior.

Muscles Affected.—*Rhomboids* present but thin. *Supra* and *Infra spinatus* more wasted than any other muscles. *Latissimus dorsi* thin and wasted below. *Pectorals* present but thin. *Deltoid* and all other muscles of arm and forearm strong and well

developed. No signs of any other affection elsewhere, excepting the *Peronei* are weak.

For about ten months she has been getting a little weak at the ankles, and she has been wearing the soles of her boots out mostly on the outer part, so much so, that she has sought medical advice for it. She can raise the foot to a right-angle, and perform all the movements of the legs strongly and well. The peronei are weak, but no marked evidence of atrophy.

Case 7.—Harry P——, aged 14. Brother to above cases.

Face Muscles Affected: *Frontales*, *Orbicularis palpebrarii*, *Corrugator supercilii*. Right *Zygomatic* and *Orbicularis oris*. The Naso labial grooves are indistinct.

Movements: Cannot frown or wrinkle the forehead. Can approximate lids, but not tightly. It is impossible for him to round the mouth as for whistling, and when he laughs the left angle of the mouth goes up and out, the right remaining stationary.

Neck and Shoulders.—Muscles affected: *Trapezius* wasted below. *Sterno cleido mastoid* thin. Slight winging of the scapulæ. A little atrophy of the *Infraspinatus*. The *Pectorals* are present. The *Deltoids*, and all other muscles of the shoulder, arm, forearm and hand, as well as those of the rest of the body, are well-developed and strong.

Case 8.—Margaret P——, age 10; sister to above cases. Fairly strong child, but has a phlyctenular ulcer on the left eye.

Face.—Muscles affected: There is not complete atrophy of any of the muscles, but she cannot wrinkle her brow, She can just move the *Corrugators supercilii*, but cannot frown. The lids can be approximated, but not tightly. She can pout the lips as for whistling, but evidently impaired a little. The *Zygomatics* act when she laughs. The *Trapezius* is thin, but all other muscles appear to be normal. There is a little winging of the scapulæ, which she can correct by muscular action.

The following is a brief summary of this family :

Husband—Unaffected.

Mrs. P——, aged 54—Facial muscles only—Zygomatics impaired.

Alice Ann P——, aged 28—Facial muscles—Zygomatics unimpaired. Slight affection of *Trapezius*.

Ellen P——, aged 25—Facial muscles to slight extent only.

Mary P——, aged 23—Facio-scapulo-humeral, well-advanced, and peronei slightly.

John P——, aged 20—Unaffected.

Elizabeth P——, aged 16—Facio-scapular and Peronei impaired at earlier stage than Mary.

Edward P——, aged 17—Very advanced Facio-scapulo-humeral.

Harry P——, aged 14—Facio-scapular at early stage—One Zygomatic not affected.

Margaret P——, aged 10—Facio-scapular at very early stage—No complete atrophy.

Remarks: In the case of John, although he can perform all movements, etc., correctly, yet there are three minor peculiarities, probably congenital.

1. He cannot speak distinctly, due to a very high and arched hard palate, and not to any muscular defect of the tongue, etc.

2. The *Sterno-cleido-mastoid* is small, and the clavicular portion thin and indistinct.

3. The *Infra spinatus* is thin, which is the more marked owing to the well-developed condition of the other scapular muscles.

Taking the following four cases in order, Margaret, Harry, Elizabeth, and Mary, we have a most instructive series from a simple impairment, through to a well marked Facio-scapulo-humeral atrophy.

Case 9.—Upper and lower extremities affected, but not the face—

In the Manchester Royal Infirmary, under the care of Dr. Dreschfeld, to whom I am indebted for allowing me to report this case, which has not been published before.

Mabel W——, age 13.

Past History.—Five years ago had scarlet fever; with this exception ailed nothing until the present illness.

Family History.—Father well; aged 40. Mother well; aged 39. Brother one; died 11 months old—bronchitis. Sisters three; quite well. Uncles and aunts well, and eight cousins healthy. No history of similar affection in family or relations.

History of Present Illness.—About two months after scarlet fever, five years ago, she began to feel weak in the right leg and ankle, and a month after in the left. Two months after, the arms began to get weak, and gradually the weakness has increased.

Present Condition, December 15th, 1893.—Looks well in face though lips project a little, and no apparent atrophy in any of the facial muscles or those of tongue, palate, pharynx or larynx. She is intelligent, and has passed the 4th Standard at school.

Muscles of Neck.—*Trapezius* wasted very much, especially below.

Appearance of Shoulders.—Bony prominences easily discerned, especially the spinatus fossæ, slight winging of the scapulæ and diplacement downwards and forwards of Acromion. *Sterno-cleido-mastoid* atrophied, especially left side; Clavicular portion only the size of a lead pencil. *Levator Anguli Scapulæ* a little wasted. *Rhomboids* very much atrophied, allowing of scapulæ leaving the chest wall, but not much winging. *Supra and Infraspinatus* much wasted, bony fossœ easily made out. *Deltoid*, absence of anterior (Clavicular), and posterior (Scapular spinal) portions, the muscle being about an inch in width, extending from Acromion to Humerus, lower part thickened on each side. *Pectorals and Latissimus dorsi* atrophied.

Arm.—The muscles of upper arm on each side are somewhat wasted, especially *Coraco-brachialis*, *Brachialis-Anticus* and *Biceps*, but *Triceps* not so much. *Supinator longus* almost dissappeared. *Forearm* and *hands* look normal.

Trunk.—*Abdominal muscles* thin. *Erector spinæ* more marked on right side than left.

Pelvic muscles.—*Psoas* and *Iliacus* weak.

Thigh.—*Glutei* look and feel full, but from their action much atrophied. *Rectus Femoris* and *Vasti* wasted, and *Hamstrings* seem somewhat atrophied. *Gracilis* and *Sartorius* are very small.

Leg.—The *Anterior Tibial* and *Peroneal muscles* are atrophied on both sides, but not much apparent wasting of leg. Calf muscles fairly developed, very little apparent atrophy. *Soleus* seems rather hard and somewhat suggestive of pseudo-hypertrophy, especially if foot is carried up to a right angle.

Foot.—Intrinsic muscles seem wasted, as bony parts are easily felt.

Movements and Resisting Powers.—Face movements seem perfect, can laugh, frown, shut lids closely, shape the mouth as for whistling, etc.

Neck.—Action of muscles weak, and apparently a great effort to raise the head from the pillow when lying in bed.

Shoulder.—Can shrug the shoulders, but this chiefly consists in rotation of the Scapulæ, owing to the upper portion of *Trapezius* pulling the Acromion upwards at each movement. Can abduct the arm, but slight resistance arrests the action. The movements are weak in all directions.

Elbow.—Flexion and extension are fairly strong.

Forearm.—Supination very weak.

Wrist.—Very strong, if hand extended and fixed, requires great force to overcome this; the same occurs at flexion of the wrist.

Hand.—If the hand is closed the fingers cannot be pulled open without great force. If extended they can be easily closed at the second and third phalangeal joints (no apparent wasting, but evidently impairment of *Lumbricales* and *Interossei*.)

Thigh.—Flexion and extension very weak.

Knee.—Extension weak.

Ankle.—The foot lies "dropped" and slightly inverted. See photo No. 15. Cannot bring the foot up to a right angle voluntarily, and cannot be carried beyond a right angle passively, owing to slight contraction of calf muscles. Resistance of *Anterior Tibial* and *Peroneal muscles* practically nil. Resistance of calf muscles great.

Position when Standing.—Marked lordosis owing to tilting of the pelvis, due to weak condition of *Glutei*. See photo No. 13.

Position when Sitting.—Owing to the pelvis resting on the Tuberosities of the Ischii it is righted and the Lordosis overcome. See Photo. No. 12. There is a little right lateral curvature of the spine.

Manner of Walking.—A combination of "high stepping," as seen in dropped feet of peripheral neuritic cases, and the oscillating movements seen in Pseudo-hypertrophic Paralysis—similar to case 3.

Manner of Getting Up From Recumbent Position.—Similar to Pseudo-hypertrophic Paralysis but not quite so far advanced as case 3. See photo No. 14.

Electrical Reaction. — No reaction of degeneration; diminished to both forms of electricity Faradic and Voltaic.

Measurements.—Around Shoulder joint, $9\frac{1}{2}$ inches, right; $9\frac{3}{4}$ inches, left; Arm, 6 inches, right; $6\frac{1}{2}$ inches, left; Forearm, $6\frac{1}{2}$ inches, right; $6\frac{3}{4}$ inches, left; Thigh, 12 inches, right; 12 inches, left; Calf, 10 inches, right; 10 inches, left.

Reflexes. *Organic.*—Normal, but latterly has desire to micturate more frequently. *Superficial*, plantar present, *Deep*, Knee jerk diminished. No ankle clonus. No fibrillar twitchings.

Sensation. Normal. No signs of Neuritis.

Summary.—Most of the voluntary muscles of the body are affected excepting those of the face, forearm, and calf. The patient does not look so thin, especially in the gluteal region, as her weakness would lead one to suspect; evidently there is increase of connective tissue in some of the muscles to account for this.

Remarks.—This is undoubtedly an example of Erb's "Juvenile" form though beginning in the legs. Among the noticeable points are:—

1. There is no family history of a similar case, all three sisters, whom I have seen, being quite healthy.

2. The onset began like many of Tooth's cases after an acute illness, and in the region supplied by the peroneal nerve; there is

no evidence of peripheral neuritis, and the atrophy seems to be purely myopathic.

3. Gluteal and some other muscles plump, though weak. In referring to Pseudo-hypertrophic Paralysis in his lectures, 1879, Gowers states at page 29, "The process which leads to enlargement and that which causes wasting may be so proportioned as to counterbalance one another, and muscles may be of normal size and yet greatly diseased. Such a condition is not rare."

4. If the foot is forcibly brought up to a right angle the calf muscles are put on the stretch, the *Gastrocnemii* seem somewhat soft, but the *Soleus* looks and feels slightly hypertrophied. Upon getting the patient to stand on the toes, she cannot jump or raise her feet together off the ground.

5. Slight affection of the muscles of the hand, similar to case five. Slight implication of the intrinsic muscles of the hand has been met with in very rare instances in Pseudo-hypertrophic cases. NOTE.—12 & 13.

6. A little irritation of the bladder.

7. Slight lateral curvature. Gowers[14] states "In all the cases I have seen the lateral curvature has been with the concavity, and spinous processes towards the right." This refers to Pseudo-hypertrophic cases.

8. Lordosis, walking, manner of getting up, etc., similar to Pseudo-hypertrophic cases.

Other interesting points are: the scapulæ are not carried away from the chest wall and vertebræ, and the acromial end does not drop nearly as much as the other cases reported. With the exception of the dropped feet, and not so much hypertrophy in the calves, she is very similar to case 11, which is one of Pseudo-hypertrophic Muscular Paralysis.

(12) Gower's Diseases of Nervous System. Vol. 1. p. 509.

(13) Sach's—New York Neurol Society, 1888.

(14) Pseudohpertrophic Muscular Paralysis. 1879. p. 32.

PSEUDO-HYPERTROPHIC MUSCULAR PARALYSIS.

The following two cases, brothers, have not been reported before:—

Case 10.—Ralph J——, age 13.

Past History.—Enjoyed good health up to about 6 years of age, when he had "slow fever;" after recovering and getting about for a month or so, he began to feel easily tired and weak in the back, rendering it difficult for him to go upstairs, and he has gradually got worse up to the present time. When 8 or 9 he noticed his calves begin to get large.

Family History.—Father healthy—no similar cases on his side. Mother healthy—had ten children, buried five: all died in infancy, excepting one who died aged 19 years, of the same complaint. Besides this patient, one brother, aged 11, is afflicted in the same way. The other three have no weakness or hypertrophy (according to the mother's statement). The baby, nearly two years old, does not walk yet. A cousin of the mother's, on the mother's side, had a boy who died of the same disease, and a sister of the mother, who has two children, boy and girl, has the former ill in a similar condition, the girl being all right.

Present Condition.—Looks well in the face, and is an intelligent lad. When stripped, the hypertrophy of his calves is very apparent. Beyond a little thinning of the posterior scapular region, and projection of vertebral border and inferior angle of scapulæ, his appearance is that of a well-developed lad. See photo No. 19.

Muscles.—*Face* muscles all right. *Trapezius* thin, disappeared at lower part. *Sterno cleido mastoid* thin, especially clavicular portion. *Levator Anguli scapulæ* present. *Rhomboideus major* thin, but present, *minor* has disappeared. *Supra spinatus* thin, but present; *Infra spinatus* thinned, a little thick and hard at central portion. *Serratus magnus* present, fibres going to lower part of scapulæ are thick. *Latissimus dorsi* wasted. *Pectoralis major* thin at upper part, disappeared at lower part. *Deltoid* very thin

at anterior and posterior portion, but middle bundle thick and hard (pseudo-hypertrophic) on both sides. *Biceps* fairly developed on each side. *Triceps* a little thickened at lower part, hard to the touch (pseudo-hypertrophic) on both sides. *Supinator longus* thin, but present on each side. *Forearm and hand* no apparent atrophy. Abdominal muscles unaffected. Back muscles somewhat atrophied. *Glutei* seem plump but hard to the touch. *Rectus femoris* thin, *Vasti* and *Adductors*, a little atrophied. Calf muscles much hypertrophied; great resistance passively but very little strength actively. *Anterior Tibial* and *Peroneal muscles* a little wasted.

Electrical Reaction.—Muscles affected are impaired to both forms of electricity, Faradic and Voltaic. No R. D. No fibrillar twitching, but a sharp tap on remaining portion of pectorals shows slight local increased irritability of the fibres.

Reflexes. *Organic*—normal. *Superficial*—plantar present. Cremasteric weak. *Deep*—knee-jerks absent. No ankle clonus.

Sensation—normal.

Measurements.—Same size on both sides: Shoulder, 10 inches. Arm, 7 inches. Forearm, 6½ inches. Middle of Thigh, 12¾ inches. Calf, 12 inches.

Movements.—Can perform all normal movements with head and upper extremities, but weakened in some of them. Can shrug shoulders, but considerable effort is required.

Arms.—Resisting power of *Deltoids* very slight, a little pressure forcing the arm down to side. *Biceps* weak. *Supinator longus* weak. *Triceps* decidedly weakened.

Forearm.—*Extensors* and *Flexors* good. *Supination* much weaker than *Pronation.*

Hand.—All movements good, but extended fingers can very easily be folded into palm from the last two joints, most marked in left hand (similar to the girl Mabel W——, Case 9.)

Back.—Weak.

Pelvis.—Can flex the thigh well, but the *Glutei* are very weak, scarcely any power in them at all.

Thigh.—*Rectus femoris* very weak.

Leg.—Patient stands with heels raised oft the ground a little, owing to the contracted calf muscles, active contraction of muscles very much impaired.

Position when Standing.—Legs apart, heels raised slightly, marked Lordosis.

Position when Sitting.—Lordosis disappears. See photo. No· 15.

Mode of Walking.—The oscillating and roll like movement, well marked, throwing all the weight over the supporting leg each time.

Mode of getting up.—The *extensors* of the knees being so weak, in order to get up from a sitting position he has to put his hands on the knees, and lean forwards, thus transferring the weight, he manages to leave the seat. After this, to gain the erect position he requires help on account of the weakness of the *Glutei* and back muscles. The photograph No. 18 shows that by climbing up the chair, at the same time lowering the *pelvis*, etc., on the principle of a lever of the first kind, the head is raised, the kyphotic-like spine is changed to one of lordosis by the weight of the *pelvis* and lower parts, and then gradually climbing higher he succeeds in getting the body, as it were, in under the head.

Case 11.—Samuel J——, aged 11, brother of above.

First began to grow weak in the back, at about six years of age. No illness preceded this.

Present condition.—Looks well in the face, and when stripped looks a fairly developed child for his age. Intelligent.

Muscles.—Face: *Naso labial* fold not present. *Zygomatics* seem deficient from their action. All other muscles present.

Neck, etc.—*Trapezius* represented by thin layer above. *Sterno mastoid* thin. *Rhomboids* atrophied. *Serratus Magnus* atrophied, thus differing from his brother. *Pectoralis* atrophied, absent at lower half. *Supraspinatus* a little atrophied. *Infraspinatus* decidedly atrophied. *Deltoid*—middle portion well developed. *Latissimus dorsi* completely atrophied. *Biceps* not much atrophied. *Triceps* slightly affected. *Supinator longus* present but thin. *Forearm* and

hand muscles seem normal. In other parts almost identical with his brother, excepting calves not so thick.

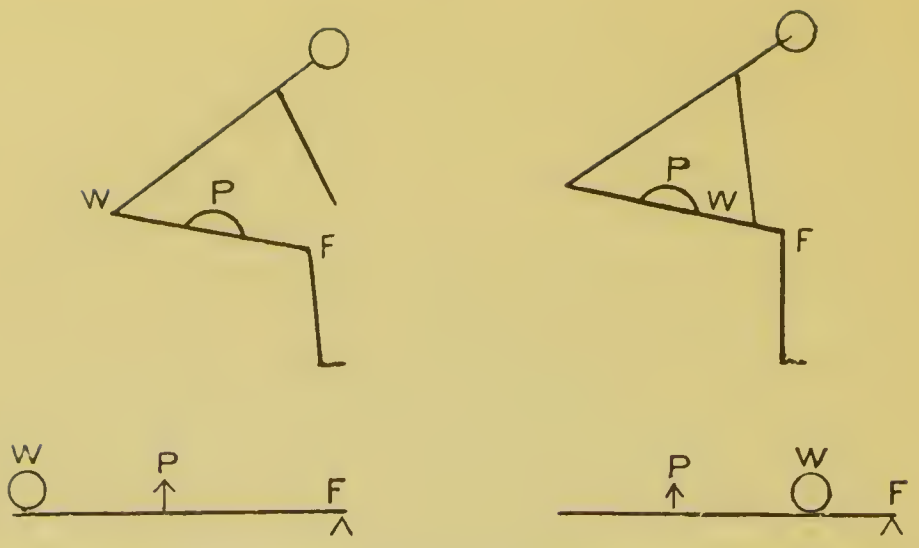
Face.—Actions normal, except that he cannot raise the angles of his mouth as in laughing.

Action of Muscles.—Not so weak as his brother. *Deltoids* have great resisting power. *Triceps* very strong. Supination a little weak. No affection of fingers.

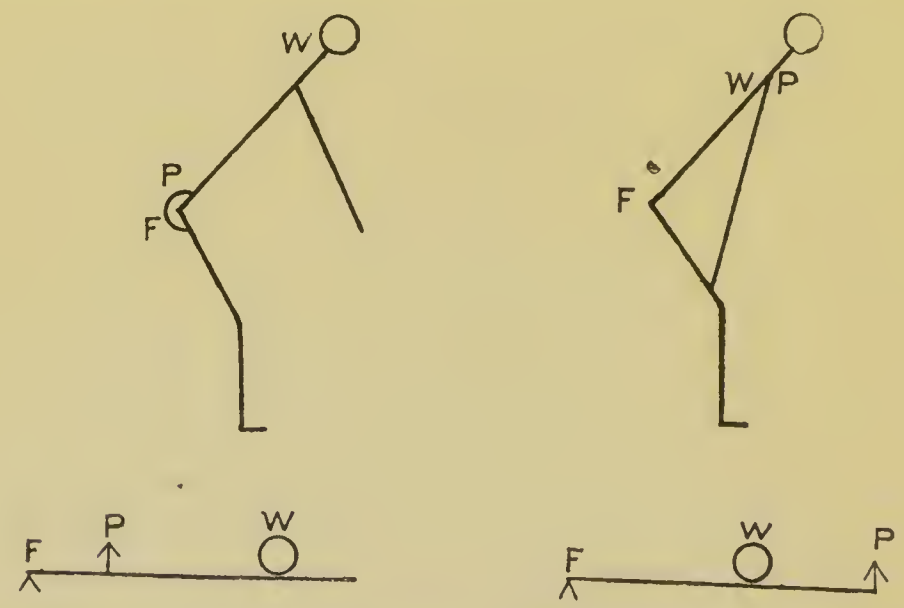
Measurements.—Shoulder, 9 inches, left; 9 inches, right. Arm 6¼ inches, left; 6¼ inches, right. Forearm, 6¼ inches, left; 6¼ inches, right. Thigh (middle), 11¾ inches, left; 12 inches, right. Calf, 10 inches, left; 9¾ inches, right.

Electrical re-action, sensation, reflexes, etc., same as brother, excepting a slight knee-jerk.

Mode of standing, walking, sitting, and getting up from the recumbent position is the same as that of his brother, except that he can rise without artificial aid. His manner of raising himself is typical, and well illustrates the two distinct actions of the hands and arms on the thighs. Appended are some snap-shots I took of him to illustrate this. As they show, the patient's first action on rising from the ground is to put his hands on his knees and lean forwards, which admits of extension of the knees. Next he grasps the thighs and gradually "climbs up" them, pushing up the body, and then, generally, the weak *Glutei* and back muscles can complete the extension of the body on the thighs. The explanation of this is that in the first action, where extension of the knee is required, we have to deal with a lever of the third kind, where the fulcrum is at the knee, the weight at the other end of the *femur*, and the mean position in which the power is applied in the middle (*Quadriceps Femoris*). By putting the hands on the knees just above the fulcrum and leaning forward, the weight is transmitted through the hands to close above the fulcrum, converting the action into that of a lever of the second kind. If the head be carried further forward and downward, the weight of this may bring the Gluteal region up without the aid of the *Quadriceps Femoris*.



Having nearly extended the thighs, the next move is to extend the trunk on the thighs. For a similar reason the weak *Glutei* cannot accomplish this and so the leverage is changed from one of the third kind to the second by the arms acting as the power,—the mean weight being then between the power and the fulcrum.



After climbing up his thighs as far as possible, as seen in photograph No. 23, the *Glutei*, *Erector Spinæ* and its continuations above must act to complete the erect posture, otherwise it is impossible, as is shown by case 10 where he cannot get into the erect position without something to lean on above the level of his

thighs. See photo No. 18. This point seems to have been overlooked in the usual descriptions.

Remarks.—These two cases show different stages of Pseudo-hypertrophic muscular paralysis; in the younger brother the *Deltoids*, *Biceps* and *Triceps* are scarcely affected; the elder brother has slight pseudo-hypertrophy of the *Deltoids*, which are very weak. The calves are also hypertropied in both, though more marked in the elder brother. The exceptional conditions of the *Zygomatics* being affected in the younger, and the impairment of the *Lumbricales* and *Interossei* of the hands of the elder brother are interesting, as similar conditions are sometimes found in simple Idiopathic Muscular Atrophy, as is seen in the cases reported in this paper.

COMPARISONS OF THE FOREGOING SIMPLE IDIOPATHIC CASES.

During the past few years the literature on this subject has considerably increased (see list in Gowers' Diseases of Nervous System, Vol. 1, p. 521), and with this a variety of divisions and sub-divisions has been given. They have been classified into: 1 The Face type. 2 Facio-Scapulo-Humeral type. 3 Erb's Juvenile type, and of the latter again sub-divisions have been made as the (*a*) Face form. (*b*) Shoulder and arm form. (*c*) Pelvic girdle form. (*d*) Leg form, etc. Although the atrophy may begin in any of the above mentioned parts, yet sooner or later, as a rule, the others get affected, and hence, the above classification is unnecessary and misleading. As a corroboration of Erb's statement that all of this variety are simply the same disease manifesting itself in different regions, we need not go out of the one family reported above for evidence. The daughters Ellen and Alice Anne, and mother present different stages of Facial affection without any other muscles being implicated, while Margaret, Harry, Elizabeth, and Mary present different stages of the Facio-Scapulo-Humeral type, and Edward shows the latter together with Pelvic girdle, Juvenile type, etc.; practically a combination of all the forms. Mabel W——, is an example of the Juvenile type, which began in the lower extremities, and has affected the pelvic girdle, and shoulders and arms, but the face has escaped. Trosier and Guinon[15] mention a case where the daughter had the Facio-Scapulo-Humeral type, the father having the shoulder and thigh muscles atrophied, but not those of the face.

Why the shoulder and arm including the *Supinator Longus* should be so commonly selected while the forearm and hand escape, as a rule, is difficult to explain.

Dr. Williamson[16] who has noted twelve cases of simple Idiopathic Muscular Atrophy in Manchester, states that eleven had the Scapulo-Humeral muscles atrophied, the odd case being Mrs. P——, recorded in this paper. Six had the face and ten the legs affected.

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(15) Rev: de Med., 1889.

(16) Medical Chronicle, Septr., 1893.

COMPARISONS OF SIMPLE IDIOPATHIC MUSCULAR ATROPHY AND PSEUDO-HYPERTROPHIC MUSCULAR PARALYSIS.

In endeavouring to arrive at a conclusion as to the similarity or difference of these two complaints, it will be necessary to consider the various details and compare them. In doing this I have chosen the following order :—

1. Appearance of patient.
2. Age.
3. Sex.
4. Heredity.
5. Causes.
6. Muscles affected.
7. Condition of affected muscles.
8. Effect of atrophied muscles.
   - (*a*) Irregular wasting.
   - (*b*) Standing.
   - (*c*) Sitting.
   - (*d*) Walking.
   - (*e*) Getting up from recumbent position.
   - (*f*) Climbing up stairs.
9. Reaction of affected muscles.
   - (*a*) Nervous.
   - (*b*) Mechanical.
   - (*c*) Electrical (Faradic, and Voltaic.)
10. Reflexes.
    - (*a*) Organic.
    - (*b*) Superficial.
    - (*c*) Deep.
11. Sensation.
12. Progress.
13. Prognosis.
14. Effect of treatment.
15. Causes of death.
16. Morbid Histology.
17. Pathology.

1. Appearance of Patient.—This varies according to the state of the disease, there may be extreme atropy as in Case 3, or apparently an almost normal condition, as in cases 1, 4, 5, 7, 8, and 9 in Idiopathic Muscular Atrophy; and in Pseudo-hypertrophic Paralysis the appearance may be normal, or with little wasting of the shoulder muscles, as in case 11, or enlargement of several muscles, particularly the calves, as in case 10.

It is interesting to compare the cases reported in this paper, as we have a varying series with well-marked Atrophy at the beginning, and a typical case of Pseudo-hypertrophy at the end. Compare the photos of Cases 3, 2, 9, 11, and 10 in the order here given :—

No. 3—Edward P——, is one of general Atrophy, with scarcely any attempt at increase of connective tissue elements.

No. 2—Mary P——, although the Atrophy is not so general, it is considerable, and there are local signs of increase of tissue in the *Infra spinatus* and lower part of *Deltoids* on each side. See photo No. 4.

No. 9—Mabel W——, in the region of *Peronei*, *Glutei*, *Arms* etc., there is no apparent wasting, and there is a little Atrophy in Scapular region and neck, but nothing like the preceding cases. These three cases are Simple Idiopathic Muscular Atrophy.

No. 11—James J——, looks in a normal condition, with the exception of a little Atrophy in the shoulder region and slight increase in the calves. This is a case of Pseudo-hypertrophic muscular paralysis.

No. 10—Ralph J——, has the calves, *Glutei*, and other muscles enlarged, and is a typical case of Pseudo-hypertrophic muscular paralysis. Apparently there is not much difference between Nos. 9 and 11 (simple Idiopathic and Pseudo-hypertrophic).

The differences between cases 1 or 2, and cases 3 and 9—all undoubtedly Simple Idiopathics—are much more than between cases 9 and 11, supposed to be different affections.

2. Age.—This has been made one of the strong points as proving a distinction between the two affections, but even admitting they appear at different ages, this does not necessarily mean the

diseases are separable. Speaking generally, Simple Idiopathic Muscular Atrophy appears later than Pseudo-hypertrophic, and the varieties of Idiopathic cases appear at different ages. According to Duchenne, the type where the face is implicated appears early, as a rule, from 5 to 7. Erb's Juvenile most commonly about puberty, etc. If, as is admitted, these varieties of Idiopathic cases appear at different ages, then it is possible on that line of argument to admit Pseudo-hypertrophic Muscular Paralysis in the same list.

There are many exceptions to the conclusions mentioned above, as for instance, Mrs. P——, aged 54, has only a few muscles of the face affected. Case 9, an example of Erb's Juvenile, began at 8. Landouzy and Déjérine have reported a case beginning at 3 years of age; on the other hand, Gowers mentions a case as late as 60. Barsickow gives a list of 17 cases where the ages were known at which the disease commenced:—One appeared at 12, four between 15 and 20, seven between 20 and 30, three between 30 and 40, and two after 40. A woman aged 52, began at 30, but her son began at 3 years of age.[17] In the article before mentioned, Erb gives the details of 23 cases of his Juvenile form; the following list shows the ages at which the disease was recognised, possibly a skilled eye would have noticed them much earlier in some cases, as it is only when there is a decided loss of use that advice is sought as a rule.

Males: 34, 20, 13, 17, 18, 6, 16, 18, 18, (Initial letter only, not certain as to sex) 40, 42, 16, 22, 12, 10—14 (brothers.) Infancy—Infancy (brothers) 14, 20, 10.

Females: 16, 10.

Pseudo-hypertrophic cases begin earlier as a rule, often the first symptoms are noticeable when the child ought to begin to walk. According to Ross, most cases begin from 5 to 13 years of age, but some are recorded where it began in adults. Ross,[18] had a case in which the patient began at 10 and was alive at 47. Gowers,[19] referring to the ages of 123 males and 16 females gives the following:

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(17) Diseases of Nervous System, Gowers' Vol. 1, p. 522.

(18) Diseases of Nervous System. Ross,—1883, Vol. 1, p. 993.

(19) Pseudo-hypertrophic Muscular Paralysis—Gowers, 1879, p. 26.

In one-third of the males the onset coincided with the first attempt at walking, half before six and three-fourths before ten. Of the girls, one case only coincided with the first walking, three before six years, and nine before ten years. Thus only 25 % of males began after ten, whilst 50 % of females began after that age. In some instances the first symptoms have presented themselves at the same age in different members of the same family. The three cases in the family reported on in this paper, all begun about six years of age, according to the mother's statement.

3. Sex.—The Simple Idiopathic variety is said to affect females, equally with males, thus differing from the Pseudo-hypertrophic which has a greater proclivity for males. As the previous list shows, this is not borne out by Erb's cases, as there are only two or three females out of 23. Webber[20] collected 41 cases of pseudo-hypertrophic paralysis, of whom five only were females. Gowers states that of 220 cases published, 190 were boys and 30 girls. The disease also appears to pursue a more chronic course in girls than in boys.[21]

4. Heredity.—The Simple Idopathic varieties are characterised by occuring often in more than one member of a family, and in more than one generation. Landouzy and Déjérine traced a group through five generations, and Barsickow reports 24 cases distributed through five generations. It is very rare for the disease to be confined to one generation.[22] The P—— family show the heredity through the mother.

Pseudo-hypertrophic paralysis is a hereditary complaint through the mother, and often, more than one member of the family becomes affected; the cases mentioned in this thesis are examples of the above. There are two mentioned by Gowers, where the fathers were affected, but there are doubts as to the correct diagnosis. There is overwhelming evidence that the inheritance is through the mother as a rule.

5. Causes.—In both Simple Idiopathic and Pseudo-hyper-

(20) Boston Medical and Surgical Journal, 1874.

(21) Diseases of Nervous system. Ross, Vol. 1, p. 993.

(22) Diseases of Nervous System, Gowers, Vol. 1, p 522.

trophic cases beyond the congenital tendency we know of no cause. Sometime a case follows on some acute illness, such as scarlet fever, measels, etc., as in case 9 (Idiopathic) and case 10 (Pseudo-hypertrophic), probably the depression of health resulting from this may have determined the time of onset, but it is not likely to have done more.

6.—Muscles Affected.—In the Simple Idiopathic variety the first to be affected as a rule are the shoulder girdle and arm; the lower part of *Trapezius*, the *Latissimus dorsi* and *Pectorals* are often completely atrophied at an early stage. Gowers says: "The tendency to atrophy of the lower part of the *pectoralis* and *latissimus dorsi* is a character which is common to this disease and pseudo-hypertrophy." Next to be affected as a rule are the *Supra* and *Infraspinatus*, *Deltoids* at times, *Biceps*, *Supinator Longus* and *Triceps*, gradually extending to others, particularly the *Glutei*, *Rectus femoris*, etc.

As a rule the muscles of the tongue, deglutition, etc., are not affected in this disease, but the following case which was observed from 1876 up to her death in 1891 suggests the possibility. It is like a case of Duchenne's Infantile form *plus* affections of the muscles mastication, etc.

Caroline Albrecht,[23] aged 32, admitted Freiburg Clinic, Dec. 22, 1876. As a school girl had nasal tone of voice, one sister and brother had similar affection of speech, slighter in degree. Parents healthy. Face rigid, mask like, Forehead smooth, *Frontales* muscles scarcely contract, *Corrugators Supercilii* contract distinctly. Insufficient closure of eyelids, marked upon the right side. Mouth widened, the right angle of mouth lower than the left. Left *naso labial* fold more marked than right, the slightly atrophied lips cannot be pointed as in whistling. *Zygomatics* and *Buccinators* act well. Speech nasal, G, K, Q and F pronounced only with great care. R not at all, B and P fairly well, D, T, L, M and N well formed, C, Z, S, Sch present some difficulty. The Tongue somewhat small, especially right half, surface smooth, no furrows, no

(23) Deutche Leitschrift, October, 1893

fibrillar jerkings. Sott palate, rises freely, but slight deviation of uvula to left during rest, and the right posterior palatial arch somewhat lower. The coarser movements of the tongue are normal, excepting a slight deviation of the apex to the right. Swallowing has to be carefully carried out, occasionally regurgitation occurs through the nose. The muscles of mastication weakly developed; frequently dislocation of jaw on both sides when the mouth is opened. Right *abductor laryngis* weak, cough hoarse, voice clear. In the neck fair sized strumous gland. Both *sterno mastoids* very thin, sternal portion of right much atrophied. *Trapezius* normal. Muscles of arm, weak, right *triceps* weaker than left. Some of the *interossei*, fourth of the left, and third upon the right, *abductor minimi digiti* upon the left side all affected. Movements in arms and hands free, co-ordination intact. Patellar reflex weak. *No sensory disturbance.*—Pupils normal, Ophthalmoscopic examination normal. No reaction of degeneration. In 1878 again admitted, *sterno mastoids* more wasted, and vocal cord paresis less marked, otherwise just the same. Tenderness on pressure over some of the peripheral nerves and at some points of the vertebral column. Occasionally polyuria up to 2,500 c. cm. In 1890 admitted again for Myoma Uteri. She died March 7th, 1891 of Purulent Bronchitis.

Post Mortem.—Nervous system intact. *Muscular System.*——All the symptoms found by Erb in Progressive Muscular Dystrophy were present. Hypervoluminous fibres not very marked; greatest width 64 micron, least 8 micron. Erb found them as high as 200 micron in some of his cases.

In the *Pseudo-hypertrophic variety* the commonest muscles to be affected with hypertrophy are the calf muscles, the disease gradually extending to identically the same muscles affected in the Simple Idiopathic variety. It is noteworthy that in many of the accounts of Simple Idiopathic cases the foot is said to be dropped and incapable of being brought up to a right angle by voluntary efforts. The calf muscles are said not to be affected as a rule. Generally the strength of muscles is tested by the resistance to pressure in their passive condition. This is not satisfactory, as the contraction of the calf muscles may afford great resistance; yet, if

ERRATUM.

Page 34, line 9. For "fair sized strumous gland" read "moderate enlargement of the Thyroid."

voluntary efforts are tried, there is an impairment of their action, as is shown, by getting the patient to raise himself on his toes as in jumping, etc. Upon comparing cases of Simple Idiopathic and Pseudo-hypertrophy, the similarity of the muscles affected is very apparent.

7. Condition of Muscles Affected.—This may be from complete atrophy of some, as in case 3, to great hypertrophy in others, as in case 10. In *Simple Idiopathic* cases the atrophy predominates, but often muscles are found with more or less increase in size, either true or false hypertrophy. Even in case 3 there is a slight attempt to hypertrophy in the *Infraspinatus* and *Deltoids*. In case 2 we find a very instructive condition, and of great importance in showing the close relationship of the two diseases. This case has been going on for 12 years; it began as a typical *Facio-scapulo-humeral* type, and although the legs are becoming affected, yet the chief mischief is confined to the neck, shoulders, and upper arms. The lower part of the *Deltoid* on each side consists of a hard mass about the size of a small hen's egg; from the weakness of action it is evidently Pseudo-hypertrophy. The *Infraspinatus* muscle is atrophied above and below, but the middle portion is enlarged, apparently beyond the normal, and this is hard to the touch.

In writing on *Pseudo-hypertrophic Paralysis*, Gowers [24] in his last edition states: "Of the condition of the muscles, that which is most characteristic is the combination of enlargement of the *Infraspinatus*, with a wasting of the *Latissimus* and lower part of the *Pectoralis*. I pointed out some years ago that this condition, which is seldom absent, is of very high diagnostic importance, and subsequent observations have fully confirmed the opinion."

In this case of *facio-scapulo-humeral* type, we have all these symptons which a careful observer like Gowers states are *characteristic* of pseudo-hypertrophic muscular paralysis. So important is this point, that he gives an illustration (figure 149, page 509, vol. 1), which is almost identical with the photograph No. 4.

(24) Diseases of Nervous System, Vol 1, p. 519.

It is interesting to note in the description of a similar case of Idiopathic atrophy, Gowers'[25] states the "*Infraspinatus* is large, but probably only hypertrophied from overuse."

In case 9, we have a condition in several muscles (*Glutei peronei arms, etc.*), where the atrophy of the muscular tissue is compensated by the increase of connective tissue elements, whilst in others the atrophy predominates.

In the pseudo-hypertrophic cases, we find all varieties, from a condition such as Duchennes "miniature Hercules" in which every muscle is enlarged, through transitions to rarer cases where all the muscles are wasted.[26] In the two cases mentioned in this paper (10 and 11) the calves vary in size, and the hypertrophy is not identical. Again looking at the cases in the order given before, we have various transitions of pseudo-hypertrophy from atrophic (case 3) through, to case 10.

8. Effects of Atrophied Muscles.—In the Simple Idiopathic variety, where there is no increase of connective tissue, some curious deformities result from the atrophy of certain muscles. The peculiar apathetic "mask like" "myopathic face," the tapir like mouth, without a smile or change of features, where all the facial muscles are affected, is very striking.

(*a*) *Irregular Wasting.* — In the case of the shoulders the acromial portion is depressed and the *scapula* looks as if slung by the *Levator Anguli Scapulæ* (see illustration Fig. 7, p. 28, Gowers, Vol. 1), and owing to gravitation, the arm is carried downwards and forwards, thus causing "Winging" of the *scapulæ.* The outer end of the clavicle is also depressed. When the *peronei* are affected, there is "dropped foot." There may be displacements and distortions of the limbs in the later stage owing to irregular contractions of the muscles. The latter condition we find most common in the later stage of pseudo-hypertrophic cases.

(*b*) *Standing.*—The patients have well marked lordosis in both Simple Idiopathic and Pseudo-hypertrophic varieties. Duchenne

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(25) Gowers' Diseases of Nervous System, Vol. 1, p 527.

(26) Pseudo-hypertrophic Muscular Paralysis, Gowers, 1879, p 30.

supposed this due to weakness of the vertebral muscles, but Gowers explains it by the weakness of the *Glutei* allowing the pelvis to rotate forward, thus carrying the Lumbar Vertebræ with it, and hence a "compensatory" curve occurs above. That this is the chief cause, is well illustrated in the photographs of cases 3 and 9 (Idiopathic) and case 10 (Pseudo-hypertrophic). See photos 5, 6, 12, 13, 16, and 19. The legs are "straddled" as a rule, and in the pseudo-hypertrophic cases the heels are slightly raised, owing to the contracted calf muscles.

(*c*) *Sitting.*—When the pelvis is resting on the tuberosities of the *Ischii* the lordosis disappears, and sometimes the curve may be in the opposite direction, well shown in both forms in photos of case 3, case 9, and case 10.

(*d*) *Walking.*—The mode of walking is similar in both Simple Idiopathic and Pseudo-hypertrophic cases where the pelvic and other muscles are affected; it is of a peculiar type, oscillating and swinging, due to throwing the weight of the body and centre of gravity each time on the leg on the ground, it has been compared to a duck waddling. Duchenne thought the oscillation of the body in walking depended upon weakness of the *Gluteus Medius*, but Ross,[27] was of the opinion it is mainly due to *contraction* of this muscle.

(*e*) *Getting up from a Recumbent Position.*—In both Simple Idiopathic and Pseudo-hypertrophic cases, the manner of getting up from the floor is identical, when the pelvic and thigh muscles are affected. Cases 3, 9, 10, and 11 all require the modifications of the levers described before—and sometimes case 9 like case 10 requires assistance to gain the erect position.—See the photos from 7 to 11; No. 14, from 16 to 19, and from 20 to 24, of cases 3, 9, 10 and 11, in the manner of getting up.

Gowers[28] on Pseudo-hypertrophic paralysis says: "The distribution of weakness in the legs causes certain peculiar defects of movement which are very characteristic, and some are even all but pathognomic of the disease."

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(27) Diseases of Nervous System. Ross, Vol 1, p. 1003.

(28) Gowers' Diseases of Nervous System, Vol. I., p 510.

(*f*) *Climbing up stairs.*—Owing to implication of the extensors of the thigh and hip, the patients in both varieties have great difficulty in going up stairs—they pull themselves up by the banisters.

9. Reaction of affected Muscles.—(*a*) *Nervous*: As long as some muscular fibres are left, the muscles can perform certain voluntary movements at the direction of the will; or act reflexly.

(*b*) *Mechanical.*—The muscles are diminished in action to a mechanical stimulus in proportion to the extent of atrophy.

(*c*) *Electrical.*—There is a diminution of power of contraction to both Faradic and Voltaic electricity in proportion to the extent of the atrophy. There is no sign of Reaction of Degeneration.—This applies to both simple Idiopathic and Pseudo-hypertrophic cases.

10. Reflexes.—(*a*) *Organic*—Swallowing, Rectal, Vesical and other organic reflexes normal—sometimes there is slight vesical irritation as in a pseudo-hypertrophic case mentioned by Gowers,[29] and in an Idiopathic case (Case 9) mentioned above.

(*b*) *Superficials*—present as a rule.

(*c*) *Deep.*—The knee jerk is often absent or impaired owing to the *Rectus femoris* being implicated. There is no ankle clonus.

11. Sensation.—This is always perfect, for touch, pain, heat, etc.

12. Progress.—In both kinds the disease varies in its course, the average time being 10 years, but in the Simple Idiopathic cases reported above, the rapidity of the disease varies in each case. Case 3 has been ill only 3 years; case 2, 12 years, and not yet so bad as case 3, etc. In Pseudo-hypertrophic cases there is the same variation, but the majority die after being affected about 10 years, between 10 and 20 years of age most commonly.

13. Prognosis.—It is doubtful whether any cases of either variety, in which the symptoms are well developed, have ever recovered; they go from bad to worse, as a rule, at a varying speed. Speaking generally, the later it appears the more slowly it advances.

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(29) Gowers' Diseases of Nervous System, Vol. 1, p 513.

14. Effect of Treatment.—Constitutional and local treatment can do nothing to stop the ultimate course of the disease, but such treatment may modify it, and a general improvement of health has been followed by slight temporary improvement in both kinds; but practically, we know of no "cure" for this affection.

15. Causes of Death.—As a rule the patients do not succumb directly to this disease, but the weakness of muscles, especially those of respiration, and impaired health, allow of the development of some intercurrent affection, often tuberculosis, which eventually carries the patient off.

16. Morbid Histology.—After most careful examinations, the conclusions arrived at by the majority of pathologists are that the chief manifestations are in the muscles themselves, the nerves, spinal cord, and brain being normal, as a rule. The changes in both Simple Idiopathic and Pseudo-hypertrophic forms are only those of degree, and microscopically they cannot be distinguished from one another. The various results are due to the double constitution of muscular tissue, the presence of interstitial connective tissue between the muscular fibres. Briefly stated there may be:

(*a*) Simple defect in the growth of the muscular fibres leading to their ultimate disappearance.

(*b*) With this simply an overgrowth of connective tissue.

(*c*) Or a perverted growth with fat cells.

Landouzy and Déjérine[30] report an Autopsy on a case of the *Facio-scapulo-humeral* type. The appearances were those of a primary degeneration of the muscles, there was very slight increase in the amount of connective tissue and fat.

In pseudo-hypertrophic paralysis we find the muscles in various stages of degeneration, and increase of connective tissue and fat cells. Gowers gives a full and lucid description.[31]

17. Pathology.—In both Idiopathic and Pseudo-hypertrophic affections the cause is due to a germinal deficiency or "congenital defect" in the muscles, potential at first, and often called out by some acute illness or exertion; always hereditary through the

(30) Revue de Médicine, 1885.

(31) Diseases of Nervous System. Vol. 1, p. 515.

mother (with doubtful exceptions mentioned before). Roth attributes it to a primary defect in the Ovum. Onanoff and others, to faults in the development of the muscle plates. Erb is inclined to attribute the changes to some affection which we cannot at present recognise, of the cells of the anterior cornua of the spinal cord, but the majority of authorities are of opinion that they are essentially a muscular disease—a true myopathic atrophy. A study of the development shows that this is possible, as the muscles are developed from the *mesoderm*, while the nervous system is derived from the *ectoderm*. Leonowa,[32] records a case of Amyelia where the muscles were perfect though there was complete absence of the Anterior cornua or nerves· In his researches he shows that striated muscular tissue is developed independently of the cells of the anterior cornua of the Cord. Ferrier,[33] sums up against the Myelopathic theory and in favour of the Myopathic.

There are differences of opinion as to how the atrophy is brought about, some contend it is due to a primary myositis and that the increased connective tissue formation and consequent atrophy of fibres constitute a later manifestation; for instance, Erb, considers the local change first appears in the muscular fibres, they hypertrophy and undergo nucleur proliferation, followed by a similar proliferation of the connective tissue elements; then follow atrophy and disappearance of the muscular fibres and a diminution in bulk of the muscle, as in Simple Idiopathic cases, or along with the atrophy of the muscular fibres there may be an exuberance of connective tissue or *lipomatosis* as in pseudo-hypertrophic cases. Roth has come to similar conclusions. These opinions were arrived at after examining portions of the affected muscles obtained by means of the "harpoon" from living subjects. Gowers considers the hypertrophy under such circumstances may be due to tetanic contraction of the fibres resulting from the irritation. But the other conditions, corroborated by the changes found after death, show that Erb's conclusions are probably correct. Others maintain the connective tissue changes are the primary cause,

(32) Neurol. Centralbl. 1893

(33) "B. M. Journal," Sept. 30th, 1893.

especially in the pseudo-hypertrophic cases. The atrophy of the muscular fibres according to this view, is the direct result of the increase of interstitial tissue. The formation of fat is merely an evidence of the lowered state of vitality of the tissues concerned, and forms no essential part of the disease.

It has been suggested that absence of the Thymus gland, or premature degeneration of this organ, may account for the disease, but feeding with minced Thymus glands has produced no good effects on such cases so far.[34]

Notwithstanding the results mentioned above, the exact primary cause of the changes in the myopathies is still doubtful. Upon comparing the cases recorded in this paper, there is much to suggest that Erb's surmises may be correct. Leaving out the affection of other parts for a time, and confining our attention to the changes in the muscles of the neck, shoulder and arm, it is striking how consistent the order is, suggesting the possibility of a central origin of the atrophy.

Ferrier and Yeo's[35] experiments on monkeys have done much to localise the groupings of muscles in the spinal cord, which, in some cases, have been corroborated clinically by Beevor.[36] Although there are differences of opinion as to the exact areas, the following may be approximately correct :—

Trapezius Sterno-mastoid, etc. = Upper cervical region.

Supra and *Infra spinatus* = As high as 4th cervical nerve (Thorburn)[37]

Scapular muscles, *Pectoralis*, *Serratus* and *Deltoid* = 5th and 6th segment (Gowers).[38] The centres for the two parts of the *Pectoralis* are separate and are associated—the *Clavicular* with that of the *Serratus magnus*, the *Costal* with that of the *Latissimus dorsi*. The *flexors* of the elbow and *Supinator longus* probably correspond nearly in level with the *Deltoid*. The *Triceps* below the above.

Ferrier,[39] in referring to Progressive Muscular Atrophy says :—

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(34) "Lancet," January 13, 1894.

(35) Proc. Royal Society, 1881, and Brain, Vol. 4, 1881.

(36) Localisation of Motor Centres in the Brachial enlargement. Med. Chir. Trans, 1885.

(37) The Surgery of the Spinal Cord.—Thorburn.

(38) Diseases of Nervous System. Vol. 1. p. 192.

(39) "British Medical Journal," Sept. 30th, 1893, p. 726.

"When the disease begins at the upper part of the cervical enlargement, the *scapular* muscles, *deltoid* and *flexors* of the arm, with the *Supinator longus* which is synergic with them, are first attacked, and the muscles of the upper arm may become entirely wasted, while those of the forearm, with the exception of the *Supinator longus*, may remain normal."

Below is a list of the cases with the names of the muscles more or less atrophied in the regions referred to.

1. Margaret P——, Impairment only.

2. Alice Ann P——, *Trapezius* only.

3. Harry P——, *Trapezius and Sterno-mastoid*, slightly affected and slight thinning of *Infra spinatus*.

4. Elizabeth P——, *Trapezius*, *Sterno-mastoid*, *Infra*, *and Supra spinatus* most wasted, *Rhomboids* thin.

5. Samuel J——, (*Pseudo-hypertrophic*), *Trapezius*, *Sterno-mastoid*, *Rhomboids*, *Supra*, *and Infra spinatus* especially thin. *Deltoids* unaffected, arm muscles and *Supinator longus* very little affected.

6. Ralph J—— (*Pseudo-hypertrophic*), *Trapezius*, *Sterno-mastoid*, *Rhomboids*, *Supra*, *and Infra spinatus*. Upper part of *Pectoralis and Serratus* present. *Deltoids* impaired, and muscles of arms, including *Supinator longus* impaired.

7. Mabel W——, *Trapezins*, *Sterno-mastoid*, *Rhomboids*, *Supra and Infra spinatus* (very thin.) *Deltoids* absent in front and behind, *Biceps* weak. *Supinator longus* wasted. *Triceps* not so much wasted.

8. Mary P——, All the muscles of the neck, shoulder, and arm including the *Supinator longus*, but escape of the forearm.

9. Edward P——, Ditto.

From the above it is evident an affection of the upper part of the cervical portion of the spinal cord, gradually extending downwards, would atrophy the muscles in exactly the order given. Whether due to some change unrecognisable microscopically, or a functional disturbance of the motorial cells of the anterior cornua of that region, is pure conjecture at present, but if due to this a similar condition of the facial nuclei in the *Medulla* might account for the atrophy of the facial muscles, and this, plus the affection of a part of the hypoglossal nucleus, would explain the unique

case reported before, where there were symptoms of Duchenne's Infantile form, and implication of the tongue and palate, but no apparent central lesion *post mortem*.

Bruce[40] shows that some fibres pass from the proximal end of the hypoglossal nucleus to the genu of the facial, and Ferrier[41] states that this may supply the *Orbicularis oris* and possibly the soft palate.

Still, after most careful search, no apparent change sufficient to account for the conditions has been found in all examinations of the cord made after death ; consequently, we must yet look upon these cases as Myopathic atrophies.

Dr. James Stewart[42] says : In the present state of our knowledge it is impossible to say whether we have to do with one disease, presenting in different cases, different anatomical features, or with a distinct series of primary muscular affections. It is contended by many that there is essentially only one form of primary muscular atrophy, and that the various clinical differences are simply accidental ; while on the other hand, it has been maintained by some, that there is a fundamental pathological difference between at least a certain number of the different types which are described. At present the weight of opinion is with the former hypothesis.

It is evident from the above comparisons that Simple Idiopathic and Pseudo-hypertrophic cases are probably different manifestations of the same disease ; for the distinction is so small that in many cases it will be impossible to discriminate between them.

From the foregoing reports it is seen that in the so-called Simple Idiopathic cases, generally some attempt at hypertrophy is found in one or more muscles ; consequently, as a comprehensive designation which will embrace all the Myopathies, that given by Erb is most suitable—" Progressive Muscular Dystrophy."

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(40) Illustrations of the mid and hind brain (Bruce.)

(41) "British Medical Journal," Sept 30th, 1893, p. 725.

(42) Keating's Diseases of Children, 1891, Vol. IV, p. 812

It is only a few years since the Atrophic variety (Simple Idiopathic) of Progressive Muscular Dystrophy, has been described separately from Progressive Muscular Atrophy. It was considered to be the latter complaint with hereditary tendencies, and occuring usually in young subjects, but there are so many points of difference that they are now considered separate diseases.

The following two cases of Progressive Muscular Atrophy are interesting, as they exhibit marked points of contrast between themselves, and are introduced here to show some of the chief differences between this disease and Progressive Muscular Dystrophy.

For the addresses of these patients I am indebted to Dr. Dreschfeld.

PROGRESSIVE MUSCULAR ATROPHY.

Case 1.—S. B——, age 62—Male. No occupation.

Past History.—Was well up to the commencement of this illness, which began about 17 years ago.

Family History.—Father and mother were healthy; both dead. Five brothers and one sister. One brother died of consumption when young. One died of an operation, age 22, and one died from some abdominal complaint, age 56. The sister is dead, from pain in the bowels (?). Two brothers alive; both healthy No history of a similar illness in any relative.

History of Present Illness.—Seventeen years ago he first noticed a wasting of the left hand, and this gradually spread up the arm. Some time after the commencement in the left hand, the right began to be affected, and he has gradually gone worse ever since.

Present Condition.—The patient stands with head bowed in front of him, and hands and arms hanging helplessly by his sides. He looks thin, but can walk and talk without much difficulty.

Face.—He has full use of his facial muscles, and there is no evidence of any impairment. The intrinsic and extrinsic muscles of the eyes are perfect. The muscles of the mouth, tongue, palate,

pharynx and larynx seem all right. He can talk fairly well, but being a Belgian, it is difficult to say if there is any impairment. His wife thinks his speech is a little thicker than it used to be.

Neck Muscles.—The *Trapezius* is very much wasted, only a few fibres at the upper part can be felt—the *ultimum moriens* of Duchenne. *Levator anguli scapulæ*, fairly well marked. *Sterno cleido mastoid* very much atrophied, only a few fibres representing the sternal portion. *Platysma myoides* well developed.

Shoulders.—*Rhomboideus major*, few fibres remaining. *Rhomboideus minor* atrophied. *Pectoralis major* atrophied. All the other shoulder muscles seem completely atrophied. See photo, No. 26.

Arms, Forearms, and Hands.—Intrinsic muscles of the *hand*, muscles of the *forearm* and *arm*, are completely atrophied on both sides; no signs of even the *Triceps*—the *ultimum moriens* of the arm.

Trunk.—*Serratus magnus* almost absent. *Latissimus dorsi* completely atrophied. *Intercostals* are thin, especially above. *Diaphragm* acting. *Erector Spinæ* and its continuations somewhat atrophied.

Pelvis.—*Glutei* atrophied as a whole, especially *Maximus* and *Minimus*; *Obturator internus*, *Pyriformis* and *Gemelli* seem affected as their movements are weak and easily resisted.

Thighs.—*Hamstrings* are a little atrophied. *Adductors*, *Sartorius*, *Rectus fermoris* and *Vasti* are wasted.

Legs.—*Peroneal* and *Anterior Tibial* muscles well developed. *Soleus* and *Gastrocnemius* fairly well developed.

Feet.—*Intrinsic* muscles of the foot are small.

Results of Atrophied Muscles. Position of the head, Flexed, but the chin does not quite touch the sternum; owing to this the spinous processes of the 7th and 8th cervical and 1st dorsal vertebræ project abnormally. The cause of this is the weakness of the *Trapezius* and other muscles going from the vertebræ to the head. This weakness is so marked, that when standing, under ordinary circumstances, he cannot raise his head, but by projecting the chest and lower part of the body, and thus altering the curvature of the spine, he succeeds in raising the head by means of the few fibres remaining, but on assuming the erect posture again, his head

drops down similar to the illustration in Gowers.[43] He can shrug his shoulders a little, chiefly by the remaining portion of *Trapezius* rotating the scapulæ. There is a little rotation of the scapulæ. See photo No. 26. *Clavicle* and *Coracoid process* are well seen.

Arms.—The Humerus on each side is rotated inwards, but the head has not apparantly left the level of the Glenoid cavity. See photo No. 25. The arms and hands are absolutely useless, they hang helplessly by his side or in front of him, with the palms backwards and thumbs inwards. There is no *main en griffe*, but as Duchenne pointed out, this is due to paralysis of the *Lumbricales* and *Interossei*, whilst the long Flexors and Extensors of the fingers act completely or partially, therefore, as all are paralysed and atrophied in this case, this symptom will naturally not present itself.

Back.—He can flex, extend, rotate and circumduct the vertebræ.

Glutei.—Can flex and extend, but evidently an exertion.

Muscles of Thigh.—Fairly strong, but slight delay in rising from sitting posture.

Calf.—*Peronei* and *Anterior Tibial* muscles all strong.

Owing to the extensive atrophy of the muscles of the neck, shoulders, and arms, the rotatory movements are all done by the vertebræ, hence the patient in getting up from a chair and turning one way or the other, looks as if supporting a head and arms not belonging to him.

Reaction.—(*a*) *Nervous.*—Where the muscles are not completely atrophied he can produce some voluntary movements.

(*b*) *Spontaneous.*—In the remaining muscles, and in fact in many regions where no muscle can be felt, there are fibrillar contractions. The *Rectus femoris*, *Peronei*, and *Calf muscles* all show it well, and according to Duchenne, this is a sign of the advancement of the disease to those parts.

(*c*) *Mechanical.*—By putting any of the muscles on the stretch the fibrillar twitching is well marked—upon bringing the head up the remaining portion of the *Sterno mastoid* shows it very well—even when passively bringing the arms away from the side a similar

(43) Diseases of Nervous System, vol. 1, p. 476, fig. 136.

fibrillar twitching in the region of the *pectorals* is seen, thus showing that every strand of muscle is not atrophied. Forcibly flexing the ankle, as in trying for ankle clonus, does not produce the latter, but many localised fibrillar contractions occur all over the muscles.

Tapping.—A sharp tap with the finger on a muscle produces a localised fibrillar contraction.

(*d*) *Thermal.*—Cold has a wonderful effect on fibrillation. Upon stripping the patient to the waist and allowing the cold air to play on him, the whole remaining muscles of his body seem to go off into a series of tetanic-like contractions. The head nods, the knees shake, and he looks as if he would totter to the ground, but all this is easily arrested by wrapping a warm rug around him.

(*e*) *Electrical.—Faradic Current.* — All the muscles remaining respond to this form of electricity, though some require a stronger current than others, and all require a slightly stronger current than normal; often the action seems delayed, and then a tonic contraction takes place in the muscle, lasting some time.

Trapezius and Sterno-cleido-mastoid require a medium strength.

Glutei require a strong current and the action is delayed.

Hamstrings similar to *Glutei.*

Rectus Femoris, Sartorius, Vasti and Adductors.—All respond to a less stimulus and give a better result.

Gastrocnemii.—Respond, but require slightly increased current.

Soleus and Flexor Longus Hallucis.—Contract to a medium current.

Peronei Ditto. —In all these the action was delayed.

Anterior Tibial.—Muscles respond better.

Interossei.—Act to a strong current.

Voltaic Current—

Trepezius, Sterno-mastoid.—Respond to a weak current, slightly weaker than is necessary to cause a contraction of my own muscles.

Glutei. Ditto.

Hamstrings Ditto.—A little more sensitive.

Rectus Femoris, Adductors and Vasti.—Require a little stronger current.

Tensor Vaginæ Femoris.— Ditto.

Calf, Anterior Tibial and Peroneal muscles require a stronger current.

Interossei and Abductor Hallucis—Ditto, and slightly delayed.

Tonic contractions resulted in some cases.

From some of the symptons of "reaction of degeneration" above, it is evident the disease is progressing; in the *Trapezius, Sterno-mastoid, Glutei, and Hamstrings* particularly.

Reflexes—(*a*) *Organic*: In swallowing a little fluid there is some delay, and a distinct double gurgle in the œsophagus, suggestive of a slight impairment, otherwise there is nothing abnormal in the act of deglutition.

No bladder or rectal trouble.

(*b*) *Superficial reflexes. Plantar* present *Cremasteric* absent.

(*c*) *Deep.*—There is no attempt at knee jerk, no ankle clonus.

Sensation.—Normal as to touch, pain, temperature, etc.

PROGRESSIVE MUSCULAR ATROPHY COMBINED WITH SPASTIC PARAPLEGIA AND BULBAR PARALYSIS.

Case 2.—Jno. Wm. C——, age 37 years. Married. Occupation, car driver.

Past History.—Never had any illness before this.

Family History. Has five brothers and five sisters, all healthy. Eight children, four dead of convulsions when very young, and four are alive and well.

History of Present Illness.—Four years ago first noticed something wrong with the fingers of the left hand, the weakness gradually spread up his arm; the right arm began some time after the left, and after working for two years longer he had to give up his employment. He has gone worse ever since.

Present Condition.—Patient looks fairly well in the face, but has a tremulous movement about the mouth, most marked on the right side and in the act of smiling. His speech is somewhat thick and hesitating, and he cannot sound some words at all clearly. The whole body and legs are in a tremulous condition from the

" fibrillar contractions," and the bed feels to vibrate in consequence.

Muscles Affected.—Those of the *mouth*, *tongue*, *palate*, *pharynx*, and muscles of *deglutition* are all affected. The intrinsic and extrinsic muscles of the eyeball are all right.

Neck.—*Trapezius* atrophied below and at middle part, the *ultimum moriens* remaining. *Sterno-cleido-mastoid*, very small bands represent the sternal and clavicular portion. *Platysma myoides* well marked. *Levator Anguli Scapulæ* not completely atrophied.

Shoulders.—*Rhomboids*, *Supra*, and *Infra Spinatus* and *Deltoid* all atrophied.

Pectoralis major.—Atrophied, but a thin portion extends from the middle of the Clavicle, and from the 4th and 5th ribs to its insertion on the Humerus, on each side.

Arms, Forearms, and Hands.—The muscles are completely atrophied, with the exception of a few fibres of the Triceps, on the left side.

Chest and Abdomen.—*Latissimus dorsi* atrophied, *Intercostals* thin, *Diaphram* acting. Abdominal muscles apparently normal.

Back.—Muscles somewhat atrophied.

Pelvis and Thigh.—Iliac fossa can be easily defined.

Glutei wasted.

Hamstrings in a tonic spasm, but knees not bent.

Rectus femoris flabby, and with marked wasting on both sides. *Vasti and Adductors* a little wasted, but in a tonic state.

Legs, etc.—*Calf muscles* well developed, no fibrillar twitchings, except when tapped. *Anterior tibial* and *Peroneal muscles* a little wasted, but no dropped ankle. All in a tonic condition.

Extensor brevis and *Abductor hallucis* seem thin. Difficult to define the other muscles of the foot.

Results of Atrophied Muscles.—*Face*.—Fibrillar contractions in *Masseters*.

Mouth —Movements are tremulous and hesitating ; cannot draw the mouth back very far ; can approximate the lips, but cannot whistle or approximate sufficiently for the production of that sound.

Tongue —The tongue lies like a flabby yellowish mass at the bottom of the mouth, with irregular coated surface. After a great

effort he can protrude it a little—about 1-16th of an inch—beyond the lower lip.

Palate.—Upon saying, "Ah," this is moved slightly upwards and to the left.

Speech.—Hesitating, thick and indistinct. Could talk perfectly well up to two years ago. "M, N, B, T, P," can be pronounced; "R and S," cannot be sounded clearly. There is a peculiar running of words together, as he says, "nine-uner-an-nine-e-nine," for nine hundred and ninety nine.

Head and Neck.—When standing, the head hangs forward, but he can raise it voluntarily, owing to the deeper muscles of the head and neck not being completely atrophied. It requires a greater effort than normal, as is shown by the Synergic contraction of the *frontalis* muscle which is associated with the extensors of the head, as in looking upwards, but should not contract when the head is raised normally. Can shrug the shoulders a little.

Hands and Arms.—Hang helplessly by his side, he can move the left elbow very slightly. No "*main en griffe*" for reasons similar to those given in the 1st case.

Back.—Although weak, he can perform the normal movements.

Pelvis and Thigh.—Can perform the normal movements though weakened.

Standing.—Can stand alone and turn around with his eyes shut.

Walking.—Stiff, spastic-like, feet coming down with heels slightly raised and a "festination" like tendency when going. Stiff and fixed at the knees.

Co-ordination.—Can co-ordinate legs and feet to a certain extent, can touch one toe on the foot with the big toe of the other foot when requested.

Reaction.—(*a*) *Nervous.*—Where the muscles are not completely atrophied, he can perform voluntary movements.

(*b*) *Spontaneous.*—The fibrillar contractions are very marked, going on continuously in one part or another, making a careful examination of the muscles almost impossible. It is very marked in the few remaining fibres of the muscles of the neck, *Pectorals*, etc.,

also very persistant in the *Rectus femoris* and *Adductors* of the thigh. Not so marked in the *Calf* and *Anterior Tibial* muscles.

(*c*) *Mechanical.*—Upon tapping, the fibrillar contractions are augmented, and well brought out in the Calf and other muscles of the leg.

(*d*) *Electrical.*—The result of this examination is unsatisfactory, as the simple application of the electrodes without any electricity caused local contractions. As far as one can judge, there seemed to be a diminution of contraction of the whole of the muscles to both Faradic and Voltaic currents, with occasional tonic contractions to Galvanism.

Reflexes. — (*a*) *Organic.* — *Respiration* normal. *Swallowing*, difficult, especially in the case of solid food. No bladder or rectal trouble.

(*b*) *Superficial.*—*Plantar* present, most marked in the left foot. *Cremasteric* present, but weak. *Abdominal* doubtful, owing to fibrillar contractions. *Epigastric* present. *Interscapular*, no muscles to respond.

(*c*) *Deep.*—*knee jerk* greatly exaggerated. An ordinary "tap" produces a clonus of about ten jerks. Upon pulling down the patella with forefinger above the superior border, and tapping, a well-marked clonus of the *Quadriceps Extensor* is obtained. *Ankle clonus* well marked on both sides, and continuous as long as hand is held against the foot. *Triceps*: On left side the reflex of Triceps is exaggerated, three or four contractions following a tap on the Triceps tendon. On the right side the atrophy of the Triceps is complete; hence no result. *Radial*: No muscles to respond.

Sensation.—Normal as to touch, pain, temperature, etc. Can distinguish two points on tongue at $\frac{1}{8}$ inch apart; $\frac{1}{4}$ inch on forehead and fingers; $1\frac{1}{2}$ inches back of forearm; 2 inches at back and chest. *Muscular sense*: Can tell the difference in weights by suspending a bag with various weights to his elevated feet and legs.

Remarks.—In the above cases the atrophy began in the intrinsic muscles of the hands and gradually extended upwards, until at present the two cases are in a somewhat similar condition as

regards the atrophy. While the first case has a history of 17 years duration, the second has one of four years only, showing the variation in rapidity of this disease. The implication of the lips, tongue, palate, etc., in the second case shows extension of the degeneration to the *Medulla*, implicating the nuclei of the Hypoglossal, etc.; the slight difficulty in swallowing in the first case is an indication of a similar extension upwards. The degeneration in the cord is also extending downwards, as is shown by the changes in the *Glutei*, *Quadriceps femoris*, and other muscles of the lower extremity.

In both cases the *ultimum moriens*, which Duchenne termed the upper part of the *Trapezius*, remains; and in the second case some fibres of the *Triceps* are present in the left arm.

Ferrier[44] has demonstrated that the various segments of the spinal cord govern different groups of muscles, apparently antagonistic in some cases, but found to work in unison to produce more or less complex movements: he thought these groupings were functional, but Sherrington,[45] Herringham, and others consider them anatomical. Each muscle is innervated from more than one group and may be connected with several, hence the brachial and other nerve plexuses are to allow of this distribution of the fibres in different trunks to the various muscles engaged in each functional combination. Beevor[46] has corroborated the above clinically; and has also shown[47] that a muscle may be paralysed to one functional combination and not to another, which may be explained by the various nerves going to different portions of the muscles as shown by Russell.[48] The above shows why the *Triceps* is the last to atrophy completely in the arm, and accounts for the persistance of the upper part of the *Trapezius*, seeing they each get their nerve supply from an extensive area.

Although there is such similarity in the atrophy, the other spinal symptoms are quite different; in the first case there is

(44) Localisation of Atrophic Paralysis, Brain, Vol. 4, p. 217.

(45) British Medical Journal, September, 30, 1893.

(46) Localisation of Motor Centres in the Brachial enlargment, Med. Chir. Trans., 1885.

(47) Brain, Volr 14, p 51.

(48) Phil. Trans.. 184 B. 1893.

absence of knee jerk, etc., while in the second this is exaggerated, and there are other evidences of *spastic paraplegia*.

The first case is very similar to those described by Aran in 1850, Cruveilhier in 1853, and Sir Wm. Roberts in 1858, and termed "protopathic" by Charcot.

The second case in some respects is similar to Charcot's description of Amyotrophic lateral sclerosis, and as he considered that in these cases atrophy is secondary, he spoke of them as "deuteropathic." Gowers[49] is of opinion that the fibres of the Pyramidal tract are more or less affected in all cases of Progressive Muscular Atrophy of spinal origin, even where there are no symptoms indicative of this. In every case where he has examined the cord *post-mortem*, he has found degeneration in the lateral tract. Strumpell[50] mentions a a case where this was not found, but there is a doubt as to the original starting point of the disease in this case.

Gowers does not attribute it to an extension laterally, but to an independent origin, *i.e.* the same condition that produces the change in the anterior cornua, also affects the lateral tract, more or less, in connection with that area. In some cases degeneration of the cortical cells of the brain and the whole of the "upper segment" has been found, but in others it has not extended above the crus; he suggests it is the perepheral portion that degenerates first; and this view seems corroborated by a case with *post-mortem* notes reported by Riley.[51] In this case the disease began in the lower part of the cord, as shown by atrophy of the muscles of the lower extremity occurring first, and gradually extending upwards until at the end of twelve months the patient died from the muscles of respiration being affected. During the progress, there were "fibrillar contractions" and other symptoms of the disease, but no increase of knee jerk, ankle clonus, or any other indication of the lateral tract being implicated; yet, *post-mortem*, degeneration of the lateral tract, as well as that of the multipolar cells of the anterior cornua was found. This was greater below where the disease

(49) Diseases of Nervous System, Vol. I. p. 489.

50) Zenkers Zeitsch, 1887, and Neur Centralbl, 1888.

51) New York Journal of Nervous and Mental Disease, 1892, p. 620.

was further advanced in the anterior cornua, and less in the cervical region, where also the cornual changes were less.

A somewhat similar case has been fully described by Dreschfeld.[52] In addition to atrophy, etc., beginning in the lower extremeties, there was a gradual loss of knee jerks. After death, great changes were found in the multipolar cells, and there was degeneration of the lateral tract, gradually diminishing higher up in the cord. This change could not be traced into the decussation.

In cases where the Spastic condition is apparent, Charcot considered the *lateral sclerosis* as the primary disease, the extension of the degeneration laterally accounting for the atrophy. Where the atrophy preceeds the spastic condition, a similar lateral extension from the anterior cornua is supposed to be the reason, according to some authorities.

Leyden suggests that the *polio-myelitis* in the cervical region is followed by secondary degeneration in the lateral tract below.

Gowers' explanation is, that if the degeneration of the cells in the anterior cornua is ahead of the degeneration in the lateral tract for any particular area, by the time the degeneration in the lateral tract has reached that region, there can be no evidence of it, owing to the reflex arc being interfered with by the atrophy of its motorial portion; but if the degeneration in the lateral tract is in advance of the changes in the anterior cornua, then evidence of this is apparent, as in case 2. In the latter case, there is no evidence from the history, that any changes began before the atrophy; there has been no rigid flexing of the arm and wrist as described by Charcot in a typical case of *Amyotrophic lateral sclerosis;* therefore, the conclusion is, that either the changes in the lateral tract took place simultaneously with those in the anterior cornua, or a little later above, but in the lower part they are in advance.

Gowers classes all these variations under the one disease, but Strumpell, although agreeing with him as to the pathological conditions, etc., yet considers the clinical features sufficiently distinct to justify separate descriptions.

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(52) Brain, 1886, Vol. VIII. p. 174.

COMPARISONS OF PROGRESSIVE MUSCULAR ATROPHY (MYELOPATHIC ORIGIN) AND PROGRESSIVE MUSCULAR DYSTROPHY (MYOPATHIC ORIGIN).

The following is a brief summary of the comparisons of the above diseases.

(1) Appearance of the Patient.—This depends on the extent and particular muscles atrophied. In advanced cases of Progressive Muscular Atrophy, the hands and arms hang helplessly by the sides, and the head is bent forward as is shown by the photos 25 and 26. If the disease begins in the upper part of the Cervical enlargement of the cord, impairment and wasting of the shoulder and arm muscles will be the first symptoms, and if the lumbar region of the cord is first affected, difficulty in walking and atrophy of the muscles in the lower extremities will be apparent.

In the myopathic varieties, the characteristic face, winging of the scapulæ, wasted appearance of the arms and normal appearance of the fore arms and hands, lordosis, peculiarity in gait, etc., and the well known symptoms of the pseudo-hypertrophic variety are distinguishing points in well marked cases, as the photographs Nos. 2 and 5, to 24, illustrate. In milder cases the local wasting may produce abnormal appearances or prevent movements, as the impossibility of quite closing the eye lids, whistling, laughing, etc., when the face is affected,—the abnormal rotation, winging of the scapulæ and atrophic appearance of the shoulder and neck when these parts are affected. In the family recorded above, it begins in the face and *Trapezius*, and extends downwards, thus differing from Progressive Muscular Atrophy which, when it does affect the upper part of the cervical portion of the cord, generally begins at the Deltoid and other upper arm and shoulder muscles, and extends upwards. When the disease affects the *Peronei* and *Anterior Tibial* muscles, there is dropped foot and high stepping walk. If the *Gluteal* region is affected, lordosis occurs, etc.

(2) Age.—As a rule, Progressive Muscular Atrophy seldom comes on before 25 years of age, generally between that and 45; while

the Dystrophies occur before that age (in childhood usually,) though there are exceptions in both diseases.

(3) Sex.—The myelopathic form is more frequent in males than females, the proportion being 3 to 1 according to Gowers. Sir Wm. Roberts found 15 women among 99 cases.

In the myopathies the atrophic variety is said to affect females equally with males, but the list given by Erb does not corroborate this: the pseudo-hypertrophic variety is commonest in males.

(4) Heredity.—As a rule Progressive Muscular Atrophy is not hereditary, sometimes there is a history of an indirect neuropathic disposition, but seldom is there a direct inheritance. Both forms of the Myopathies are hereditary, and several may be affected in a family, as the examples given in this thesis prove. When many members of a family suffer from muscular atrophy, the malady is generally idiopathic and not spinal.[53]

(5) Occupation.—It has been suggested that over-use of the muscles, leading to exhaustion, may account for Progressive Muscular Atrophy, but there is no history of such in either of the above cases. The social condition seems to have no marked influence on the Dystrophies, and there is no evidence that overwork produces the disease, though the use of the muscles, or some acute illness, may show up the defect.

(6) Causes.—What the direct causes are in both these affections are doubtful. Exposure to cold, exhaustion, syphilis, etc., have been credited with causing the Myelopathic form, but the majority have no such history. The primary cause of the atrophy in this disease is due to degeneration of the multipolar cells in the anterior coruna of the spinal cord, while in the Dystrophies, the supposed cause is the "germinal defect" of the muscles themselves.

(7) Muscles affected.—In the Myelopathic variety, the Atrophy most frequently begins in the intrinsic muscles of one hand, often the thumb muscles first, soon followed by a similar condition in the other hand, and a gradual extension of the changes

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(53) Disease of Nervous System—Gowers, vol. 1, p. 472

up the limb in order of the groupings in the segments of the spinal cord. Occasionally the shoulder group, or that of the lower extremity, may first be attacked. The face is seldom affected, but in advanced cases the muscles of the mouth, tongue, palate, and deglutition may be affected, owing to an extension of the disease up the cord to the Medulla, as in the second case described above. More rarely it has begun in this region.

In the Dystrophic varieties the facial muscles are often affected, as the reports above show; very seldom are those of the tongue or palate atrophied, but one case has been reported and is referred to in this paper. The muscles of the hand and forearm are seldom affected until late in the disease, thus contrasting strongly with Progressive Muscular Atrophy. In the case of Edward P——, when examined again on February 12th, 1894, the atrophy was advancing in the forearms, the *Extensors Carpi Radialis Longior* and *Brevior* being much atrophied, and there was a diminution in size of all the muscles of the forearm and hand in comparison with their condition two months previously. In the Pseudo-hypertrophic variety the *calf muscles*, *Glutei*, and *Shoulder muscles* are the most commonly affected.

8. Condition of Affected Muscles. — As the atrophy advances there is sometimes an increase of connective or fatty tissue in Progressive Muscular Atrophy. As before described, among the Dystrophies all conditions from atrophy to pseudo-hypertrophy may be found.

9. Effects of Atrophy.—This depends on the muscles affected. In the Progressive Muscular Atrophy of the usual type, the useless arms and dropped head already referred to, and in advanced cases the difficulty in talking, etc., when the *Medulla* is affected are distinguishing points; while among the Dystrophies the commonest results of atrophy are shown by the Facial expression when the muscles are affected, winging of the *scapulæ lodorsis*, dropped feet, peculiarities in gait, and in getting up from a recumbent position, etc., already described; and the enlarged calves and other muscles, with marked weakness in the pseudo-hypertrophic varieties, and in advanced cases of the latter disease,

kyphosis, and other deformities resulting from the irregular wasting of the muscles.

10. Reaction of Muscles. (*a*) *Nervous.*—Some voluntary movements can be produced in both varieties, provided there are a sufficient number of healthy fibres left in a muscle to contract.

(*b*) *Spontaneous.*—A very important condition is seen in both the cases of Myelopathic origin described above, namely, spontaneous fibrillar contractions of the muscles, increased by allowing the cold air to play on the skin. This at one time was thought "diagnostic" of this complaint, but cases of this disease have been recorded without these symptoms, and although rare, they have occurred in some examples of the Dystrophies.[54]

(*c*) *Mechanical.*—Similar local contractions are readily brought out by tapping the muscles with the fingers in Progressive Muscular Atrophy, but not as a rule in the Dystrophies, yet in the case of Harry P——, where the *Pectorals* are getting thin, a local contraction is obtained on tapping with the finger, and the same result is obtainable in the second pseudo-hypertrophic case. This symptom is seen in other diseases, especially those of a wasting character, such as phthisis, etc.

(*d*) *Electrical.*—There is a difference in the reaction of the two diseases as a rule.

In Progressive Muscular Atrophy, there is often a modified R.D. There may be a gradual diminution to both currents when transmitted through the nerves, but the muscles may give a prolonged contraction to the galvanic current, and may respond as readily to this current as under normal conditions, and occasionally even more so. Or the A.C.C. may equal K.C.C. There may be the qualitative change of R.D., without the Voltaic increase. Among the Dystrophies there is a gradual loss of reaction to both currents in proportion to the atrophy—no signs of R D., but Tooth[54] states this has been recorded in connection with a case of Myopathic Atrophy.

11. Reflexes.—In the protopathic form of Progressive Muscular Atrophy, there is a gradual loss of all reflexes excepting the organic,

(54) St. Bartholomew's Hospital Reports, 1889, p. 143.

and that may be affected late in the disease, as the interference with swallowing etc., shows.

In Amyotrophic lateral sclerosis, there is an exaggeration of the deep reflexes with ankle clonus, etc., and the organic reflex of swallowing, etc., may be affected early.

In the Dystrophies, there is a gradual loss of reflexes, superficial and deep. The organic reflexes are normal as a rule, but one case where swallowing, etc., was interfered with, is referred to in this thesis.

12. Sensation.—Normal in both forms as a rule, though localised areas of anasthesia have been recorded in some cases of Progressive Muscular Atrophy.

13. Progress.—Variable in both diseases. May be completely arrested, but this is very rare.

14. Prognosis.—Grave in both.

15. Effects of Treatment.—Gowers[55] states that Hypodermic injections of small doses of strychnine will sometimes arrest Progressive Muscular Atrophy Duchenne[56] quoted cases which were improved by faradism, while Remak thought galvanism of value. The late Dr. Ross had great confidence in the latter, but Gowers considers it useless. In the case of Dystrophies, no cure has been found. Cases are sometimes improved for a time by attention to the general rules of health.

16. Causes of Death.—In both diseases, generally some intercurrent affection is the immediate cause of death. Sometimes it is due to extension of the atrophy to the muscles of respiration as in Riley's case before mentioned.

17. Morbid Histology.—In Progressive Muscular Atrophy, besides the changes found in the muscles, there are evidences of the disease found in the nerves, nerve roots, and spinal cord. As far back as 1853, Cruveilhier found the anterior roots of the spinal nerves were grey and atrophied in a *post mortem* examination made on Lecomte, a celebrated rope dancer of that period, who died

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(55) Diseases of the Nervous System, Vol 1, p. 496.

(56) Fagge's Medicine, Vol 1, p. 506.

with symptoms of this disease. Lockhart Smith first discovered the changes in the spinal cord, but Charcot was the first to associate the disease with changes in the anterior cornual cells. The following is a brief summary of the changes found in this disease :—

(*a*) *Muscles.*—In the same muscle may be found healthy fibres, and others in all stages of degeneration, fatty and vitreous. There is an increase of connective tissue elements as seen by the greater number of nuclei, formation of connective tissue, and occasionally an increase of fat.

(*b*) *Nerves.*—Degeneration of some fibres, which if traced up are found to come from the anterior cornua.

(*c*) *Nerve roots.*—Degeneration of many of the fibres of the anterior roots, the posterior remaining normal.

(*d*) *Spinal Cord.—Grey matter.*—Degenerative changes in the anterior cornua, and sometimes anterior commisure. Diminution, pigmentation, vacuolation, and disappearance of some of the multipolar cells in the anterior cornua, and shrivelling or disappearance of their processes. Increase of connective tissue elements (Deiters cells, etc.) These changes are found greatest in extent corresponding with the nerves to the most affected parts. The posterior part of the grey matter is normal.

White portion.—The direct and crossed pyramidal tracts are generally found degenerated.

In the Progressive Muscular Dystrophies, the changes before described are noticed in the muscles, but the Nerves, Nerve roots, and Spinal Cord are usually found unaffected.

According to Erb, in the early stages there is a true hypertrophy of some of the muscular fibres, a condition thought to be characteristic of the muscular dystrophies in distinction from the spinal atrophies. Besides this swelling and hypertrophy, there is atrophy well marked in other fibres; the bundles are rounded, there is increase of nuclei, splitting of fibres, vacuolization, and tendency to break up into fibrilliæ. Next, the connective tissue elements increase, and this is followed by further atrophy of the muscular fibres, with increase of connective tissue, until in some cases, a dense, hard, myo-sclerosis results, and there may be a deposit of fat in the

connective tissue cells, etc. Occasionally there is a secondary degeneration in the terminal nerve filaments.[5]

18. Pathology.—In Progressive Muscular Atrophy, Duchenne thought the wasting of the muscles was due to an alteration of the multipolar cells in their *trophic* action, and that the nervous *motor* action was not interfered with. Erb has shown that this erroneous conclusion was due to the reaction of the degenerated fibres being concealed by the healthy fibres surrounding them. According to Gowers [58]:—"The essential lesion of the disease is a slow decay of the lower segment of the motor path—the segment which consists of the ganglion cells and their prolongations in the axis cylinders of the nerve fibres. To this the conspicuous symptom—the muscular wasting—is secondary."

The views regarding the changes in the lateral tract have been referred to above.

As before mentioned, the Progressive Muscular Dystrophies are supposed to be due to the defects in the muscles themselves, but the late Dr. Fagge [59] was strongly inclined to the opinion, that pseudo-hypertrophic paralysis will ultimately be found to be a spinal affection, a view which is held by Erb at the present time concerning all the dystrophies. Dreschfeld [60] has reported a case of Erb's Juvenile paralysis in which the patient died, aged 42, in the Manchester Royal Infirmary, December, 1884. Microscopic examination of the cord and nerves showed very little abnormal changes. In the cord a few of the motor ganglion cells appeared smaller, and the processes shrivelled. There was an increase of Deiter's cells. The nerves were apparently normal. Atrophic changes were found in the muscles; the author concludes by stating,—"The appearance of the muscles would lead one to think that the atrophy is secondary."

If the disease is of primary muscular origin, it is difficult to explain such a regularity in the affection of the muscles as found in

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(57) Text Book of Nervous Diseases—Dana, 1892, p, 272.

(58) Diseases of Nervous System, Vol. I., p. 491.

(59) Fagge's "Medicine," Vol. 1., p 518.

(60) Brain, vol, 9, p. 194.

eight cases of the atrophic form in one family, and two of the pseudo-hypertrophic form in another family described in this thesis. Upon studying the original derivation of the various muscles, the degeneration does not occur as one would expect, if of purely muscular origin. Considering the muscles entending from the trunk to the arm, we find, that originally, as each limb begins, bud like, to project from the side of the embryo, it receives an enveloping sheath of muscle from the side of the body, at first as a funnel-like mass which grows in connection with the limb; eventually the upper portion becomes differentiated into an anterior and posterior series of muscles, passing from the trunk to the shoulder girdle and humerus. Anteriorly are the *Pectorals* and *Subclavius*, and posteriorly a superior and inferior group. The superior mass includes a superficial sheet consisting of the *Cleido Mastoid* and *Trapezius*, the latter of which is continued to the humerus by the *Deltoid*, and a deeper sheet constituted by the *Rhomboids* and *Serratus Magnus*, with its cervical prolongation, the *Levator Anguli Scapulæ*. The inferior mass is composed of the *Latissimus dorsi*, and a deeper part in the *Teres Major*. All the other muscles of the limb are intrinsic.[61]

Seeing that the *Deltoid* is practically a prolongation of the *Trapezius*, if there is a primary defect in the latter, one would expect a similar change in the former, and if so, they should degenerate together when there is this defect; but in all the cases recorded, the *Deltoid* is either not affected, or, if so, it occurs later in the disease when the arm muscles are beginning to atrophy. The *Latissimus dorsi* disappears early in the disease, but the *Teres major* later. As all the muscles of the limb, excepting those enumerated above are intrinsic, it is remarkable that the atrophy, if it is not of spinal origin, has such a sharp line of demarcation as a rule, as the arm muscles and *Supinator longus* in the upper extremity.

One of the strong reasons brought forward by Ferrier at the meeting of the British Medical Association, August, 1893, against classing these diseases as myelopathic, was, that the *Deltoid* is seldom affected when the other muscles of the shoulder are

(61) Quain's Anatomy, Vol. II. Part II. p. 274, 1892.

atrophied. According to the groupings in the cord the *Deltoid* is situated as low as the flexors of the arm (Gowers), and it is remarkable that in all the cases mentioned in this paper, when the arm muscles are affected the *Deltoid* is also.

Out of 12 cases of the atrophic variety seen by Dr. Williamson,[62] 7 had the *Deltoids* affected. In the report of Dr. Dreschfeld's case referred to above, it is stated:—The *Biceps*, *Triceps*, and *Supinator longus* were very much atrophied. The *Deltoid* muscle was impaired, and the patient was not able to hold his arm horizontally for more than a minute or two.

I have confined the descriptions to the cervical portion of the cord and its corresponding muscles, as the other portions of the cord have not been so fully worked out, but the order of affection in the lower extremities is very similar to the groupings suggested by Ferrier for the lumbar region of the cord. Ferrier,[63] at the conclusion of his discourse on Myopathic Atrophy remarks:—The whole subject is still in need of further elucidation.

The following are, therefore, the chief differences from a *Clinical* point of view.

1. *Appearance.*—The disfigurements from the atrophy, already described.

2. *Age.*—The Myelopathies occur after 30, the Myopathies before 20, as a rule.

3. *Heredity.*—Seldom in Myelopathies, common in Myopathies.

4. *Muscles affected.*—Generally begins in the small muscles of the hand in myelopathic atrophy; but in the myopathic variety, anywhere except in the small hand-muscles, which are usually respected to the last.

5. *Effects of Atrophy.*—Position of arms, head, lordosis, peculiarities in gait, etc., already described.

6. *Reaction of Muscles.*—Spontaneous fibrillar contractions common in Progressive Muscular Atrophy, very uncommon in the

(62) "Medical Chronicle," Sept. 1893

(63) "British Medical Journal," Sept. 30th, 1893.

myopathies. *Electrical*—modified R.D., common in later stage of myelopathies. As a rule no R.D. in myopathies.

7. *Reflexes.*— Deep, increased in the Amyotrophic lateral sclerosis variety. Generally diminished or lost in the others.

EXPLANATION OF PHOTOGRAPHS.

SIMPLE IDIOPATHIC MUSCULAR ATROPHY.

No. 1.—Mrs. P——. Action of Muscles when endeavouring to say "O."

No. 2.—Mary P——. Cannot approximate the eye lids.
No. 3.— Do. Displacement of Clavicle, etc.
No. 4.— Do. Back view, showing pseudo-hypertrophy of *Infra spinatus* and *Deltoid* (lower part) with the absence of *Rhomboids*, *Latissimus dorsi*, etc., on each side.

No. 5.—Edward P——. "Tapir like" mouth. Lordosis. Tilting of pelvis. Winging of scapulæ. Marked atrophy, etc.
No. 6.— Do. When sitting, disappearance of lordosis.
No. 7, No. 8, No. 9, No. 10, No. 11 — Do. Manner of gaining the erect position from the floor.

No. 12.—Mabel W——. Sitting. No lordosis. Atrophy of the *Infra spinatus*, etc.
No. 13.— Do. Standing. Lordosis, etc.
No. 14.— Do. Manner of gaining the erect position.
No. 15.— Do. Dropped feet from affection of the *Peronei* and *Anterior tibial* muscles.

PSEUDO-HYPERTROPHIC MUSCULAR PARALYSIS.

No. 16.—Ralph J——. Absence of lordosis when sitting. Pseudo-hypertrophy of calves, etc.

No. 17.— Do. Manner of getting up from a seat.

No. 18.— Do. Manner of gaining erect position.

No. 19.— Do. Standing. Lordosis, etc.

No. 20, No. 21, No. 22, No. 23, No. 24 — Samuel J——. Manner of gaining erect position from the floor, illustrating the different applications of leverages explained in the text. Compare with Edward P—— above.

PROGRESSIVE MUSCULAR ATROPHY.

No. 25.—S. B——. Front view, showing atrophy of arm, etc. Clavicle, coracoid process, and internal rotation of the head of the humerus well shown.

No. 26.— Do. Back view, showing atrophy of the muscles of the arms, scapulæ, etc.


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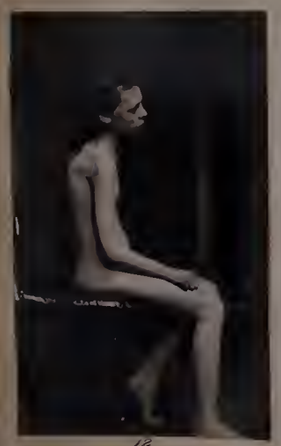
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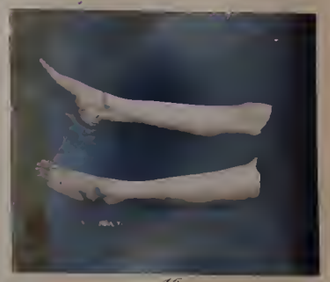
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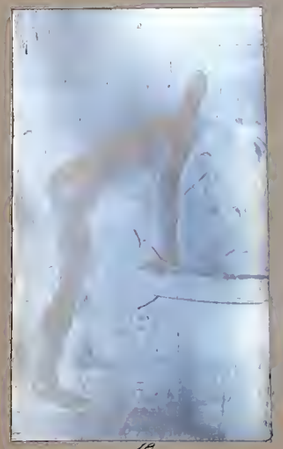
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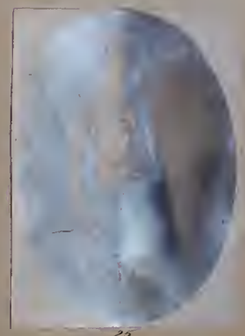
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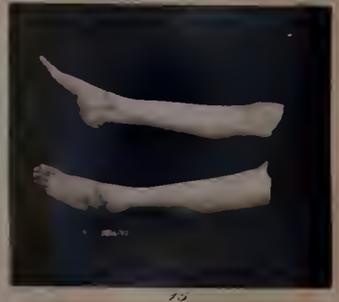

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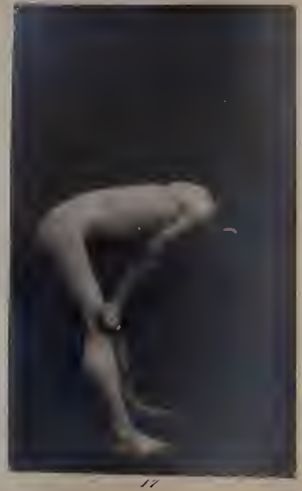

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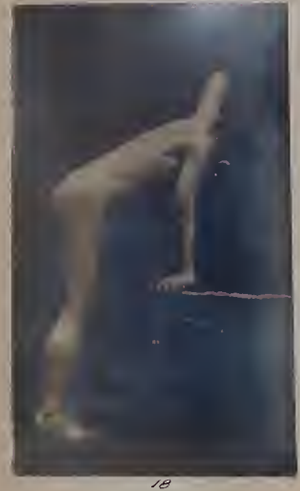

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